Tumoral, quasitumoral and pseudotumoral lesions of the superficial and somatic soft tissue: new entities and new variants of old entities recorded during the last 25 years. Part XII: Appendix

Lesioni tumorali, quasitumorali e pseudotumorali delle parti molli somatiche e superficiali: entità nuove e varianti di entità già note, descritte negli ultimi 25 anni. Parte XII: Appendix

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Summary
In an eleven part series published in Pathologica, we have presented various tumoral, quasitumoral and pseudotumoral lesions of the superficial and somatic soft tissue (ST), which emerged as new entities or as variants of established entities during the last quarter of a century. Detailed clinicomorphological and differential diagnostic features of approximately sixty entities were chosen on the basis of their clinical significance and morphologic distinctiveness. The series included fibrous and myofibroblastic tumors (e.g. solitary fibrous tumor, high grade classic and pigmented dermatofibrosarcoma protuberosans, inflammatory myofibroblastic tumor and myofibrosarcomas), fibromyxoid and fibrohistiocytic neoplasms (e.g. Evans’ tumor, phosphaturic mesenchymal tumor, inflammatory myxohyaline tumor), special adipocytic / vascular / and smooth muscle lesions (e.g. chondroid lipoma, Dabska’s tumor, ST hemangioblastoma, lipoleiomyosarcoma), epithelioid mesenchymal malignancies of diverse lineages (e.g. epithelioid liposarcoma, proximal-type epithelioid sarcoma, neuroendocrine extraskeletal chordoid myxoid sarcoma), ST Ewing’s tumor and peripheral nerve sheath tumors (perineuriomas and pigmented and rosetting tumors of the schwannoma/neurofibroma group), extranodal dendritic or histiocytic proliferative processes (follicular dendritic cell sarcoma, Rosai-Dorfman disease, Castleman’s disease, and pleomorphic xanthomatosus tumor), and tumors with myoepithelial differentiation. The section devoted to selected pseudotumoral entities included representatives of the hamartoma type (neural fibrolipomatous hamartoma, ectopic hamartomatous thymoma, rudimentary meningocele), metabolic diseases (amyloid tumor, nephrogenic fibrosing dermopathy, tophaceous pseudogout, pseudoinfiltrative parathyromatosis), stromal tissue reactions to trauma (fibroosseous pseudotumors of digits) and infections (bacillary angiomatosis), and normal organs (glomus coccygeum).

To conclude the descriptive phase, supplementary material has now been collected and appended in an attempt to provide a quick digest of essential knowledge both for comparison and differential diagnosis. The data have been tailored to synthesize diverse sources, integrating clinical elements and references to articles that previously appeared in Part I (“Introduction”), Part II (“The List and Review of New Entities”) and Parts III to XI (“Excerpta”). At the very least we hope this final part (“Appendix”) will provide the reader with a useful tabular organization of ST lesions and a reference resource.

Introduction
In 2003 Dr Bisceglia was commissioned by the Editors of Pathologica to produce a series of articles concerning tumoral, quasitumoral or pseudotumoral lesions of superficial and somatic soft tissue recognised in the last quarter century, including newly described entities and new variants of established entities. Through a series of 11 articles published in the journal 1-11, Dr Bisceglia and colleagues have presented both a global overview of the numerous entities which have been described in this period, with few exceptions dating from 1979, and also...
provided detailed discussion, including any novel data, of selected entities. This information was presented in a logical and algorithmic manner, both in extensive tabular and discursive formats, supported by a comprehensive but selected, and up-to-date bibliography. This allows the reader to navigate through the vast amount of information which has accrued in the area of soft tissue pathology in the last 25 years or so. The intent, and its desired result, is that this treatise should be a valuable resource not only for pathologists who regularly encounter soft tissue lesions in their practice, but more importantly for those who navigate the diagnostic soft tissue traps on a more sporadic basis, for residents and fellows in histopathology, and clinicians dealing with soft tissue lesions. A brief summary of the approach used in this series of articles is presented here to facilitate retrospective navigation through the series.

The first article (in Italian) provides an introduction and raison d'être for the project, criteria for categorization and a detailed reasoning for inclusion or exclusion of various conditions. It ends with a “master table” of diagnostic categories, presented in 2 broad groupings of “Tumoral and Quasitumoral Lesions” (18 diagnostic categories) and “Pseudotumoral Lesions” (10 diagnostic categories). This Table forms the backbone for the subsequent articles and excerpta which expand in detail on many, but not all lesions which are included in each diagnostic category. To compile the comprehensive catalogue of lesions considered in the subsequent articles in this series, the authors have drawn not only on an exhaustive literature review (which includes peer-reviewed articles and important discussions appearing as letters to the Editor), but also the considerable personal experience of many of them in the field of soft tissue pathology. The authors have wherever possible listed entities according to their presumed or accepted histogenesis/differentiation, while a category is retained for those of uncertain histogenesis/differentiation. The rationale for certain morphological groupings is provided, for example, those defined on the basis of a distinctive extracellular matrix (e.g. fibromyxoid tumors), or cellular composition (e.g. fibrohistiocytic or fibromyofibroblastic tumors).

The second article, prefaced with a preamble in Italian, presents the comprehensive list of entities (in English) which fall within each diagnostic category, accompanied by key references. The latter include important references of a general or review nature in respect of particular diagnostic categories/entities, in addition to references specific to particular entities and their variants. While all of the diagnostic categories of the 2002 WHO classification of soft tissue tumors are included in the master table, by necessity and design, even within the categories of tumorous lesions, there has been a deliberate “splitting” so that in contrast to the WHO scheme, some categories appear in their own right as major diagnostic categories, e.g. the myoepithelial tumors, malignant mixed mesenchymal tumors and neoplasms of perivascular epithelioid cell origin. It is the authors’ intention that the “exploded” list of entities with their pertinent bibliography can provide the casual reader, as well as a pathologist dealing with a specific soft tissue problem, an intuitive and rapid means of assessing the breadth of differential diagnostic possibilities and the state of knowledge that exists at the time of publication. Selected entities to be discussed in clinicopathologic detail and illustrated in the subsequent articles (“Excerpta”) are highlighted in the list. They were selected for detailed discussion on the basis of their clinical importance and/or their distinct morphological features.

The third to eleventh articles in the series (Excerpta 1 to 9) are devoted to presenting the clinicopathologic and biologic features of specific entities (58 in all) in detail. The following Table summarizes the entities which are discussed in depth in each Excerpta.
<table>
<thead>
<tr>
<th>Series article #/ Ref. #</th>
<th>Entities presented in detail</th>
<th>Author (initials)</th>
<th>Pathologica fascicle</th>
</tr>
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<tbody>
<tr>
<td>Part 3 / Excerpta 1</td>
<td>1. Dermatofibrosacoma protuberans, classic, and pigmented sarcomatous</td>
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<td>GIUGNO</td>
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<tr>
<td></td>
<td>2. Atypical fibroxanthoma, non-pleomorphic spindle-cell, and aneurysmal (pigmented)</td>
<td>MB</td>
<td>(June)</td>
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<tr>
<td></td>
<td>3. Plexiform fibrohistiocytic tumor</td>
<td>KC</td>
<td>2004</td>
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<td></td>
<td>4. Hemosiderotic fibrohistiocytic lipomatous lesion</td>
<td>DVK</td>
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<td></td>
<td>5. Nuchal-type fibroma</td>
<td>MM</td>
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<td>6. Desmoplastic fibroblastoma</td>
<td>CF</td>
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<td>7. Solitary fibrous tumor</td>
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<td>8. Pleomorphic hyalinizing angiectatic tumor</td>
<td>SS</td>
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<td>9. Sclerosing epithelioid fibrosarcoma</td>
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<td>Part 4 / Excerpta 2</td>
<td>1. Dermatomyofibroma</td>
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<td>2. Palisaded myofibroblastoma / intranodal hemorrhagic spindle cell tumor with &quot;amianthoid&quot; fibres</td>
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<tr>
<td></td>
<td>3. Myofibrosarcoma, low grade and high grade</td>
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<td>2004</td>
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<tr>
<td></td>
<td>4. Angiomyofibroblastoma of vulva</td>
<td>DVK</td>
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<td>5. Lipomatous hemangiopericytoma</td>
<td>CF</td>
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<td>6. Inflammatory pseudotumor / inflammatory myofibroblastic tumor</td>
<td>MB</td>
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<td></td>
<td>7. Calcifying fibrous (pseudotumor)</td>
<td>MB</td>
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<td></td>
<td>8. Inflammatory myxohyaline tumor / myxoinflammatory fibroblastic sarcoma</td>
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<td></td>
<td>9. Superficial angiomyxoma and juxtarticular myxoma</td>
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<td></td>
<td>10. Ossifying fibromyxoid tumor</td>
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<td>11. Low grade fibromyxoid sarcoma &amp; hylalising spindle cell tumor with giant rosettes</td>
<td>KC</td>
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<td>Part 5 / Excerpta 3</td>
<td>1. Phosphaturic mesenchymal tumor</td>
<td>MB</td>
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<td>2. Chondroid lipoma</td>
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<td>3. Dendritic fibromyxolipoma</td>
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<td>4. Epithelioid liposarcoma</td>
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<td>5. Lipoleiomyosarcoma</td>
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<td>6. Epithelioid leiomyosarcoma</td>
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<td>7. Granular cell leiomyosarcoma</td>
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<td>Part 6 / Excerpta 4</td>
<td>1. Acquired elastic hemangioma</td>
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<td>2. Epithelioid hemangoendothelioma of soft tissue and skin</td>
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<td></td>
<td>3. Extraneuraxial hemangioendothelioma of soft tissue and skin</td>
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<td>4. Dabska's tumor arising in lymphangioma</td>
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<td>Part 7 / Excerpta 5</td>
<td>1. Psammomatous melanotic Schwannoma</td>
<td>KC</td>
<td>APRILE</td>
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<td></td>
<td>2. Pigmented neurofibroma</td>
<td>MM</td>
<td>(April)</td>
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<td></td>
<td>3. Dendritic cell neurofibroma with pseudorosettes</td>
<td>DVK</td>
<td>2005</td>
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<td></td>
<td>4. Epithelial sheath neurroma</td>
<td>DVK</td>
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<tr>
<td></td>
<td>5. Perineuriomas - intraneural and extraneural forms</td>
<td>MB</td>
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<td>6. Ewing's sarcoma of the skin</td>
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<tr>
<td>Part 8 / Excerpta 6</td>
<td>1. Extraskeletal myxoid chondrosarcoma with neuroendocrine differentiation</td>
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<tr>
<td></td>
<td>2. Proximal epithelioid sarcoma</td>
<td>CF</td>
<td>(June)</td>
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<td>3. Extra-renal malignant rhabdoid tumor</td>
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<td>4. Follicular dendritic cell sarcoma</td>
<td>CF</td>
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<td>5. Castleman's disease of the subcutis and skeletal muscle</td>
<td>DVK</td>
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<td>6. Rosai-Dorfman disease of soft tissue</td>
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<td>7. Plexiform xanthomatosus tumor</td>
<td>MM</td>
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<td>Part 9 / Excerpta 7</td>
<td>1. Hyaline cell-rich chondroid syringoma and myoepithelial tumors - mixed tumor and myoepithelioma</td>
<td>MB</td>
<td>OTTOBRE</td>
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<tr>
<td></td>
<td>2. Dabska's parachordoma</td>
<td>MM</td>
<td>(October)</td>
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<tr>
<td></td>
<td>3. Oncocytoma</td>
<td>MB</td>
<td>2005</td>
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<tr>
<td>Part 10 / Excerpta 8</td>
<td>1. Ectopic hamartomatous thymoma</td>
<td>DVK</td>
<td>DICEMBRE</td>
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<tr>
<td></td>
<td>2. Hamartoma of the scalp with ectopic meningothelial elements / rudimentary meningiocele</td>
<td>DVK</td>
<td>(December)</td>
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<td>3. Neural fibrolipomatous hamartoma</td>
<td>MB</td>
<td>2005</td>
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<td>4. Ischemic fascitis (atypical decubital fibroplasia)</td>
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<td>5. Nephrogenic fibrosing dermatopathy</td>
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<td>6. Basalcell epithelioid angiomatosis</td>
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<td>Part 11 / Excerpta 9</td>
<td>1. Fibro-osseous pseudotumor of the digits and bizarre parosteal osteochondromatous proliferation (Nora's lesion)</td>
<td>MB</td>
<td>GIUGNO</td>
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<td></td>
<td>2. Tophaceous pseudogout (calcium pyrophosphate dihydrate crystals deposition disease – tumoral form)</td>
<td>MB</td>
<td>(June)</td>
</tr>
<tr>
<td></td>
<td>3. Amyloid tumor of soft tissue and breast</td>
<td>MB</td>
<td>2006</td>
</tr>
<tr>
<td></td>
<td>4. Gliomus coccygeum</td>
<td>MB</td>
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<tr>
<td></td>
<td>5. Post-implant parathyromatosis</td>
<td>MB</td>
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</table>
In this final APPENDIX (Part XII) to the series, 30 “problem-oriented” Tables (with bibliography) are provided to aid the decision making process in everyday diagnostic practice in soft tissue pathology. These Tables provide a comprehensive, though admittedly not exhaustive grouping of lesions according to important clues based on cytoarchitectural pattern recognition, and also in consideration of particular clinical circumstances. The Tables should serve not only as an aide-de-memoir but also as a diagnostic “check-list” when confronted with a particular diagnostic problem. To provide maximum benefit from this problem-oriented tabular approach, we thought that it would be useful to broaden the scope of the tables beyond those lesions affecting superficial and somatic soft tissues, as was our original intent in the series, to also include lesions affecting other anatomic parts/organs, e.g. the body cavities. However, genuine soft tissue visceral organ-based lesions have been excluded from the analysis. Updated references for various entities considered in the preceding Excerpta are provided, in addition to important reviews and references of a more generic nature. References to the new entities which had been already quoted in the General List of entities (Part II) or were detailed in the Excerpta – with very few exceptions limited to Table 30 – do not appear in this Appendix, to avoid repetition. Entities which have been previously described or referenced are indicated by means of various typographical options.

The Key to navigating the Tables is as follows.

• **Bolded** entities appeared in Excerpta and are listed alphabetically in the Index at the end of this Appendix, accompanied by key references that have appeared since the publication of the excerpta.

• **Italicized** entities were listed in Part II of the Series with their annotated bibliography.

• **Referenced** entities in the Tables correspond to recent articles that appeared after Part II of our series was published, or were not included in the previous List, or are considered to be key and/or review articles.

To keep abreast of the burgeoning growth in knowledge of soft tissue tumefactive conditions can be overwhelming, and to mine the breadth and depth of the data base excerpted from the eleven preceding publications requires an agility that can only be met by the power of online search engines. Since the publication of the series in Pathologica, outstanding contributions have appeared in the world literature. In this Appendix we broaden our previous list of entities and update the bibliography. To complement the traditional morphologic approach used throughout the series, the reader is referred to the January 2006 issue of Histopathology, which is devoted to soft tissue tumors and which highlights the increasing role of new diagnostic modalities, such as genetic analysis and microarray technology, in addition to the traditional contributions of immunohistochemical and ultrastructural examination. Continued refinement in nosology is possible through advances in the expertise of the investigator and the diagnostic tools being applied. Histological classifications and subclassifications that are practically achieved, reproducible and uniquely predictive, either alone or in conjunction with other patient’s characteristics, are valuable in the design and evaluation of therapeutic regimens. It is our hope that the series with its comprehensive Appendix will provide the reader with a rapid reference base, maximum availability, and practical usefulness.
“Problem-oriented” tables

With the intent to allow one, at a glance, to be pointed in the right direction, and hopefully provoke a highly satisfying “Eureka” moment!!

1. Richly collagenized lesions
   A. Tumor-like lesions
   B. Tumoral lesions

2. Myxoid lesions
   A. Myxomatous lesions
   B. Occasionally myxomatous lesions, or with myxomatous change

3. Tumors and pseudotumors with calcifications, or with chondro-osseous metaplasia
   A. Dystrophic and psammomatous calcifications
   B. Chondroid and osseous metaplasia

4. Tumors and pseudotumors with inflammatory cell component
   A. Lymphoid infiltrate
   B. Xanthomatos infiltrate

5. Atypical / bizarre / multinucleated / pleomorphic giant cell-containing tumors and pseudotumors
   A. Atypical / bizarre / monster cells in benign lesions
   B. Reactive multinucleated, osteoclast-like giant cells in benign tumors and pseudotumors
   C. Osteoclast-like giant cells in borderline and malignant tumors
   D. Multinucleated giant cells (intrinsici) in benign tumors
   E. Neoplastic giant cells (intrinsic or accompanying) in borderline and malignant tumors
   F. “Malignant fibrous histiocytoma”-like pattern

6. Tumors and pseudotumors with a distinctive architecture / growth pattern
   A. Fascicular pattern
   B. Storiform pattern
   C. Whorled pattern
   D. Palisading pattern
   E. Amianthoid structures and (pseudo-)rosettes
   F. Cording / chordoid pattern
   G. Endocrinoid pattern
   H. Alveolar pattern
   I. Pseudoglandular pattern
   J. Pseudovascular pattern
   K. Retiform pattern
   L. Papillary pattern

7. Distinctive vascular patterned lesions (non-endothelial)
   A. Hemangiopericytoma-like pattern as a constant feature
   B. Hemangiopericytoma-like pattern as an occasional feature
   C. Perivascular hyalinization prominent
   D. Plexiform capillary pattern
   E. Glomeruloid capillary pattern
   F. Hemorrhagic changes / pattern
   G. Aneurysmal changes / pattern

8. Combined, polyphasic, and heterologous tumors (other than collision tumors)
   A. Combined (homologous and heterologous) tumors
   B. Biphasic homologous (bimorphic) tumors
   C. Biphasic heterologous tumors – with epithelial elements
   D. Pseudo-biphasic tumors
   E. Biphasic tumors with skeletal muscle differentiation

9. Histogenetically diverse but morphologically similar tumors and pseudotumors with ....
   A. Round-cell / epithelioid / plasmacytoid features
   B. Clear cell changes
   C. (Uni-/multi-)vacuolated cells, (pseudo-)lipoblasts, and (pseudo-)physalipherous cells
   D. Eosinophilic / oncocytic features
   E. Granular cells
   F. Rhabdoid / pseudo-rhabdoid features (extra-renal)
   G. Ganglion / ganglion-like cells
   H. Small round cells
   I. Pleomorphic cells
10. Tumors and pseudotumors with diagnostic clues
   A. Architecturally distinct features
   B. Stromal peculiarities
   C. Cyto-histologically distinct features

11. Histologically pigmented tumors and pseudotumors with ....
   A. Melanin
   B. Hemosiderin
   C. Carbon

12. Clinically alarming benign tumors and pseudotumors
   A. Multiple, multicentric, recurring benign lesions
   B. Painful soft tissue tumors and pseudotumors

13. Pathologically alarming benign tumors and pseudotumors
   A. Multilobular / mosaic growth pattern
   B. Plexiform, infiltrating margins
   C. Intravascular growth
   D. Hypercellular benign lesions

14. Histologically benign lesions mistaken for malignant
   A. Tumor-like lesions
   B. Tumoral lesions

15. Reactive endothelial lesions that may histologically mimic malignancy

16. Histologically malignant tumors mistaken for benign lesions

17. Atypical variants of typical tumors
   A. Clinically atypical
   B. Behaviourally atypical
   C. Macroscopically atypical
   D. Morphologically atypical (histologic/cytologic)

18. Cystic tumors and pseudotumors
   A. Primarily cystic
   B. Secondarily cystic or with microcystic change
   C. With pseudocystic pattern

19. Acral tumors and pseudotumors

20. Orbital tumors and pseudotumors

21. Paratesticular tumors and pseudotumors
   A. Benign tumors and pseudotumors
   B. Malignant tumors

22. Vulvo-vaginal tumors and pseudotumors

23. Aerodigestive tract lesions (involving the orifices)
   A. Nasal cavity
   B. Oral cavity
   C. Anal canal

24. Intranodal primary tumors and pseudotumors (other than hematolymphoid)

25. Mammary stromal spindle cell tumors and tumor-like lesions and mimics
   A. Monomorphic spindle cell tumors of specialized mammary stroma
   B. Monomorphic spindle cell tumors of non-specialized mammary stroma
   C. Spindle cell myoepithelioma
   D. Spindle cell epithelial tumors

26. Infantile-juvenile tumoral and pseudotumoral lesions
   A. Hamartomas – Developmental lesions
   B. Benign tumors and pseudotumors
   C. Intermediate and malignant tumors

27. Lesions as sentinel of clinical entities

28. Lesions as sentinel of clinical syndromes

29. Embryological rests or normal anatomic structures potentially mistaken for tumors

30. Grade-based categorization of soft tissue tumors (of relevance to management)
Table I. Richly collagenized lesions

A. Tumor-like lesions
- Fibrous papule of the face (nose) (classic type and variants) 21
- Multiple perifollicular fibroma of the face and neck
- Angiofibromas (associated and unassociated with tuberous sclerosis complex)
- Collagenoma (connective tissue nevus)
- Rudimentary supernumerary digit
- Acquired digital fibrokeratoma
- Sclerotic fibroma (circumscribed storiform collagenoma)
- Reactive fibrosis
- Hypertrophic scar & keloid
- Knuckle pads
- Diabetic scleredema (upper back)
- Elastofibroma
- Infantile digital fibromatosis
- Gingival fibromatosis
- Juvenile hyaline fibromatosis 22
- Fibrous proliferations in Proteus syndrome
- Intra-articular nodular fasciitis 23
- Calcifying fibrous pseudotumor
- Idiopathic retroperitoneal fibrosis (Ormond’s disease)
- Reactive nodular fibrous pseudotumors (of the gastrointestinal tract and mesentery) 24, 25
- Sclerosing mediastinitis 25a
- Sclerosing extramedullary hematopoietic tumor

B. Tumoral lesions
- Nasopharyngeal angiofibroma
- Sclerosing/desmoplastic melanocytic nevi (ordinary acquired melanocytic nevus, Spitz nevus, blue nevus)
- Dermatofibroma variant (keloidal, sclerotic)
- Pleomorphic fibroma
- Nuchal-type fibroma (of nuchal and extra-nuchal sites)
- Gardner-associated fibroma
- Fibroma of tendon sheath
- Giant cell fibroblastoma
- Sclerotic lipoma
- Fibrous spindle cell lipoma
- Sclerosing neurofibroma
- Desmoplastic cutaneous ganglioneuroma
- Atypical fibroxanthoma with prominent sclerosis
- Tenosynovial giant cell tumor – sclerotic variant
- Desmoplastic fibroblastoma (collagenous fibroma)
- Sclerosing perineurioma
- Superficial (fascial) fibromatosis (palmar / plantar / penile types)
- Deep (musculoaponeurotic) fibromatosis (desmoid tumors: extraabdominal / abdominal / intraabdominal types)
- Paratesticular fibroma
- Solitary fibrous tumor 26
- Epithelioid hemangioendothelioma
- Sclerosing epithelioid fibrosarcoma
- Sclerosing primary cutaneous leiomyosarcoma
- “Fibroma-like” epithelioid sarcoma (- desmoplastic variant) 26a
- Sclerosing rhabdomyosarcoma
- Sclerosing paragangioma 27
- Monophasic synovial sarcoma
- Leiomyomatosis peritonealis disseminata (end-stage)
- Desmoplastic diffuse mesothelioma
- Desmoplastic small round cell tumor
- Desmoplastic melanoma 27a
- Lymphoma (occasional)
### Table II. Myxoid lesions

#### A. Myxomatous lesions

- **Focal mucinosis (cutaneous and subcutaneous)**
- **Polyvinylpyrrolidone granuloma** *(rare: may occur in the skin/subcutis/soft tissue/bone)*
- **Superficial angiomyxoma (cutaneous and subcutaneous myxoma) & juxta-articular myxoma**
- **Neurothekeoma, classic or myxoid type**
- **Nerve sheath myxoma** *(‘perineurial myxoma’ of Harkin and Reed – “plexiform myxoma”)*
- **Myxolipoma (myxoid lipoma)**
- **Angiomyxolipoma**
- **Dendritic fibromyxolipoma** / **Myxoid spindle cell lipoma**
- **Intramural myxoma**
- **Myxopapillary ependymoma**
- **Superficial acral fibromyxoma**
- **Aggressive angiomyxoma**
- **Ossifying fibromyxoid tumor**
- **Low-grade fibromyxoid sarcoma (Evans’ tumor)**
- **Superficial low-grade fibromyxoid sarcoma (Evans’ tumor)**
- **Primitive myxoid mesenchymal tumor of infancy**
- **Myxofibrosarcoma (low grade myxoid malignant fibrous histiocytoma)**
- **Extraskelatal myxoid chondrosarcoma (chordoid sarcoma)**
- **Extraskeletal myxoid chondrosarcoma with neuroendocrine differentiation**
- **Myxoid liposarcoma**
- **Primary heterotopic chordoma** / **chordoma periphericum**
- **Parachordoma**
- **Mixed tumors of salivary glands (recurrence in soft tissue)**
- **Metastatic myxoid sarcomas to soft tissue and skin**
- **Metastatic chordoma**
- **Metastatic colloid carcinoma**
- **Pseudomyxoma peritonei**

#### B. Lesions occasionally myxomatous or with myxomatous change

- **Trichodiscoma (fibrofolliculoma)**
- **Keloid**
- **Nodular fasciitis**
- **Infantile fibromatosis**
- **Subungual pleomorphic fibroma – myxoid variant**
- **Proliferative funiculitis**
- **Fibrous umbilical polyp**
- **Cutaneous myxoid fibroblastoma**
- **Giant cell fibroblastoma**
- **Dermatofibrosarcoma protuberans – myxoid variant**
- **Myoepithelioma**
- **Lipoblastoma**
- **Leiomyoma**
- **Schwannoma – myxoid variant**
- **Neurofibroma – myxoid variant**
- **Dendrocytoma – myxoid variant**
- **Chondroma – myxoid variant**
- **Fetal-type rhabdomyoma**
- **Spindle cell lipoma (occasional)**
- **Perineurioma** – **myxoid variant**
- **Dermatofibrosarcoma protuberans – myxoid variant**
- **Solitary fibrous tumor – myxoid variant**
- **Leiomyosarcoma – myxoid variant**
- **Malignant peripheral nerve sheath tumor**
- **Rhabdomyosarcoma (mainly botryoid embryonal type)**
- **Inflammatory acral myxo-hyaline fibroblastic sarcoma**
- **(Extra-)gastrointestinal stromal tumor**
- **Fibrous synovial sarcoma**
- **Clear cell sarcoma – myxoid variant**
- **Lymphoma (occasional)**
- **Melanoma (primary, and metastatic) – myxomatous** [and pseudomyxomatous variants]
### Table III. Tumors and pseudo-tumors with calcifications, or with chondro-osseous metaplasia.

**A. Tumors and pseudo-tumors with dystrophic and psammomatous calcifications**

- Hemangiomas (phleboliths mainly in cavernous and spindle cell hemangioma)
- Cutaneous meningioma
- Amyloid tumor (even bone)
- Tophaceous gout (even bone)
- Tophaceous pseudogout (tumoral crystal calcium pyrophosphate dihydrate deposition disease)
- Malakoplakia
- Fat necrosis
- Calcificion – several types
- Calcifying aponeurotic fibroma
- Myofibroma/myofibromatosis
- Calcifying fibrous pseudotumors
- Leiomyoma
- Neurothekeoma
- Ancient schwannoma
- Phosphaturic mesenchymal tumor (mixed connective tissue variant)
- Hyalinizing spindle cell tumor with giant rosettes
- Ganglioneuroma
- Psammomatous melanotic schwannoma
- Psammomatous malignant melanoma
- Epithelioid sarcoma
- Mesothelioma
- Synovial sarcoma
- Differentiating neuroblastoma
- Alveolar soft part sarcoma with psammomatous bodies, and calcifications (occasional)
- Any sarcoma with preceding necrosis (may undergo dystrophic calcification)
- Extragonadal (retroperitoneal) burnt-out germ cell tumors

**B. Soft tissue tumors and pseudotumors with chondroid & osseous metaplasia**

- (Pilomatrixoma)
- Ectopic ossification of the auricle secondary to frostbite
- Nuchal fibrocartilaginous pseudotumor
- Heterotopic chondro-ossification
- Chondro-mesenchymal hamartoma of the nose
- Mesenchymal hamartoma of the chest wall
- Fibrochondroid hamartoma
- Rhabdomyomatous mesenchymal hamartoma of the skin (occasional)
- Hyperplastic callus in pathologic conditions (e.g. osteogenesis imperfecta, ...)
- Tumefactive soft tissue extension of Paget's pseudosarcoma
- Cavernous hemangioma with ossification (occasional)
- Dermatofibroma (ossifying variant)
- Benign fibrous histiocytoma of subcutaneous and deep soft tissue (occasional)
- Fibromatosis (mainly palmar type)
- Juvenile xanthogranuloma (1 case with ossification)
- Myofibromatosis
- Epithelioid hemangioendothelioma with ossification
- Cutaneous chondroma (solitary form, multiple / familial form)
- Cutaneous osteoma
- Chondro-osteo-lipoma / benign mesenchymoma
- Hyaline cell-rich chondroid syringoma
- Chondroid lipoma
- Soft tissue chondroma & periosteal chondroma
- Extra-articular synovial chondromatosis
- Nodular fasciitis, mainly parosteal variant (fasciitis ossificans)
- Myositis & panniculitis ossificans
- Heterotopic mesenteric ossification
- Subungual (Dupuytren's) exostosis / turret exostosis (acquired osteochondroma) of the hands and feet
- Fibroosseous pseudotumor of the digits (florid reactive periostitis of the tubular bones of the hands and feet)
- Florid reactive periostitis of the long bones (e.g. tibia)
- Bizarre parosteal osteochondromatous proliferation of the hands and feet (Nora's lesion)
- Bizarre parosteal osteochondromatous proliferation of the long bones (Nora's lesion)
- Fibrodysplasia ossificans progressiva
- Ossified meningioma
- Ossifying fibromyxoid tumor
- Atypical fibroxanthoma with osteoid production
- Atypical fibroxanthoma with chondroid differentiation

**Mixed tumor of the skin and soft tissue**
- Giant cell tumor of tendon sheath & giant cell tumor of soft tissue
- Phosphaturic mesenchymal tumor (mixed connective tissue variant)
- Hemangiopericytoma
- Schwannoma with bone or cartilage
- Malignant peripheral nerve sheath tumor
- Malignant fibrous histiocytoma
- Epithelioid sarcoma (with chondroid and osseous metaplasia)
- Myxoid liposarcoma
- Embryonal rhabdomyosarcoma
- Extraskeletal myxoid chondrosarcoma (chordoid sarcoma)
- Synovial sarcoma
- Extraskeletal soft tissue & synovial chondrosarcoma
- Soft tissue mesenchymal chondrosarcoma
- Perosteal chondrosarcoma
- Parosteal, periosseous, high-grade surface osteosarcoma
- Soft tissue extraskeletal osteosarcoma
- Primary cutaneous osteosarcoma
- Primary osteogenic sarcoma of the penis
- Primary osteogenic sarcoma of the breast
- Malignant mesenchymoma
- Mesothelioma (occasional)
- Extraskeletal PNET (with massive osteocartilaginous differentiation)
- Melanoma (osteogenic melanoma)
- Metastatic metaplastic breast carcinoma, malignant mixed mullerian tumor, skeletal and extraskeletal osteogenic sarcoma
Table IV. Tumors and pseudotumors with inflammatory cell component.

A. Tumors and pseudotumors with lymphoid infiltrate
- Inflammatory fibrous papule of the face
- Lymphangioma
- Amyloid tumor
- Atypical mycobacterial spindle cell pseudotumor
- Pseudoneoplastic (histoid) leprosy
- Malakoplakia
- Bacillary angiomatosis
- Angiolymphoedematous hyperplasia with eosinophilia
- Kimura’s disease
- Lymphangioma-like Kaposis sarcoma
- “Lymphocyte-rich hobnail hemangioendothelioma” (including retiform hemangioendothelioma and Dabska’s tumor)
- Rosai-Dorfman disease, extranodal
  - Reactive nodular fibrous pseudotumors (of the gastrointestinal tract and mesentery)
  - Sclerosing mesenteritis, mesenteric panniculitis, mesenteric lipodystrophy, and other inflammatory pseudotumors
  - Sclerosing retroperitonitis (Ormond’s disease)
  - Sclerosing mediastinitis (idiopathic fibroinflammatory lesions of the mediastinum)
- Inflammatory pseudotumor (inflammatory myofibroblastic tumor)
  - “Inflammatory fibrosarcoma”
  - Inflammatory liposarcoma (lymphocyte-rich well differentiated liposarcoma)
  - Inflammatory leiomyosarcoma
  - Inflammatory malignant fibrous histiocytoma / undifferentiated pleomorphic sarcoma with prominent inflammation
  - Malignant fibrous histiocytoma – pseudolymphomatous variant
  - Angiomatoid fibrous histiocytoma
  - Acral inflammatory myxohyaline tumor (myxoinflammatory fibroblastic sarcoma)
  - Dedifferentiated liposarcoma – pseudolymphomatous variant
  - Diffuse-type giant cell tumor
  - Mesothelioma (lymphohistiocytoid variant)
  - Follicular dendritic cell sarcoma

B. Tumors and tumor-like lesions with xanthomatous infiltrate
- Xanthomatous leprosy
- Atypical mycobacteriosis
- Nodular panniculitis
- Nodular-cystic encapsulated fat necrosis (“encapsulated mobile lipoma”)
- Paraffinoma / oleogranuloma (other than lung paraffinoma and coeliac lipogranulomatous lymphadenitis)
- Lipoma with fat necrosis
- Juvenile xanthogranuloma
- Necrobiotic xanthogranuloma
- Xanthomatous / cholesterolotic benign fibrous histiocytoma
- Xanthoma (tuberous type)
- Plexiform xanthomatous tumor
- Xanthogranulomatous inflammation NOS (of any site, including abdomen and pelvis)
- Pelvic xanthogranulomatous inflammation of infective etiology – overt (e.g. actinomycoses) or occult
- Tamman-Horsfall protein deposition (early phases)
- Erdheim-Chester disease with xanthogranulomatous inflammation
- Erdheim-Chester of the breast
- Tenosynovial giant cell tumor both localized and diffuse-type
- Schwannoma
- Malignant fibrous histiocytoma – inflammatory type
Table V. Atypical / bizarre / multinucleated / pleomorphic giant cell-containing tumors and pseudotumors.

A. Benign tumors with atypical / bizarre / monster cells
- Pleomorphic fibroma of the skin
- Subungual pleomorphic fibroma
- Pleomorphic fibroma of tendon sheath
- Pleomorphic lipoma
- Pleomorphic leiomyoma
- Atypical decubital fibroplasia of debilitated people (ischemic fasciitis)
- Orbital & extraorbital giant cell angiofibroblastoma
- Pseudosarcomatous benign fibrous histiocytoma
- Atypical lipoma
- Ancient schwannoma
- Paragangioma
- Neurofibroma with nuclear atypia
- Symplastic glomus tumor
- Symplastic leiomyoma
- Bizarre angioleiomyoma
- Atrophic striated muscle cells in several conditions
- Regenerating skeletal muscle
- Pseudosarcomatous polyp
- Cellular pseudosarcomatous fibroepithelial stromal polyp of the lower female genital tract

B. Benign tumors and pseudotumors with reactive multinucleated, osteoclast-like giant cells
- Suture granuloma
- Textilomas \(^{51}\) and teflonomas \(^{52}\)
- Paraffinoma / oleogranuloma / silicone granuloma
- Amyloid tumor
- Fat necrosis
- Tumoral calcinosis
- Tophaceous gout
- Tophaceous pseudogout (nodular deposition of calcium pyrophosphate dihydrate crystals)
- Dermatofibroma with osteoclast-like giant cells
- Benign fibrous histiocytoma of subcutaneous and deep soft tissue
- Fibroma of tendon sheath (variant)
- Myofibroma / myofibromatosis (rare)
- Fibromatosis (plantar type)
- Giant cell collagenoma
- Soft tissue aneurysmal bone cyst
- Giant cell tumor of tendon sheath (localized-type) \(^{53}\)
- Chondroma (calcified variant)
- Giant cell tumor of the skin
- Giant cell tumor of soft tissue
- Giant cell tumor of mammary stroma
- Calcifying aponeurotic fibroma
- Florid reactive periostitis (fibroosseous pseudotumor of the digits)
- Extraskeletal chondroma
- Aggressive angiomyxoma (occasional)

C. Borderline & malignant tumors with osteoclast-like giant cells
- Atypical fibroxanthoma with osteoclast-like giant cells
- Plexiform fibrohistiocytic tumor
- Poshphaturic mesenchymal tumor (mixed connective tissue variant)
- Soft tissue giant cell tumor
- Giant cell tumor of tendon sheath (diffuse type)
- Leiomyosarcoma with osteoclastic-like giant cells
- Malignant peripheral nerve sheath tumor with osteoclast-like giant cells
- Extraskeletal osteoclastic (giant cell-rich) osteosarcoma
- Osteoclastic giant cell malignant fibrous histiocytoma
- Undifferentiated pleomorphic sarcoma with osteoclastic giant cells
### D. Benign tumors with intrinsic multinucleated giant cells
- Juvenile xanthogranuloma (Touton cells)
- Multinucleated giant cells angiohistiocytoma of the skin
- Kimura’s disease (polykaryocytes)
- Langerhans’ cell histiocytosis
- Myelolipoma (megakaryocytes)
- Sclerosing extramedullary hematopoietic tumor (megakaryocytes)
- Orbital and extra-orbital giant cell angiofibroma (relevant to solitary fibrous tumor group)
- Giant cell fibroblastoma
- Giant cell angiofibroma of Nakagawa

### E. Borderline and malignant tumors with either intrinsic or accompanying neoplastic giant cells
- Plexiform fibrohistiocytic tumor
- Pleomorphic hyalinizing angiectatic tumor
- Inflammatory myxohyaline tumor (inflammatory fibroblastic sarcoma)
- Giant cell angiofibroma (of orbital and extra-orbital sites)
- Malignant fibrous histiocytoma – storiform-pleomorphic type
- Malignant fibrous histiocytoma – giant cell type
- Myxoid malignant fibrous histiocytoma (gr. II)
- Clear cell sarcoma
- Alveolar rhabdomyosarcoma with pleomorphic giant cells
- Angiomatoid fibrous histiocytoma with pleomorphic giant cells
- Aggressive angiomyxoma
- Alveolar soft part sarcoma
- Soft tissue osteosarcoma
- Malignant mesenchymoma
- Melanoma (other than pleomorphic) 54
- Epithelioid atypical angiomyxoid fibromatoses
- Any pleomorphic sarcoma (pleomorphic leiomyosarcoma, pleomorphic rhabdomyosarcoma, pleomorphic liposarcoma, undifferentiated high grade pleomorphic sarcoma, and others)

### F. Malignant tumors with “malignant fibrous histiocytoma”-like pattern
- Malignant fibrous histiocytoma
- Dermatofibrosarcoma protuberans with transformation to malignant fibrous histiocytoma
- Dedifferentiated MFH-like areas in liposarcoma
- Inflammatory and pleomorphic leiomyosarcoma
- Malignant mesenchymoma with multinucleated osteoclastic-like cells
- Malignant melanoma 54
- Metastatic sarcomatoid/metaplastic carcinoma (e.g. renal cell carcinoma)
Table VI. Tumors/pseudotumors with a distinctive architecture / growth pattern.

A. Tumors and pseudotumors with fascicular pattern
- Pseudoneoplastic leprosy (histoid variety)
- Atypical mycobacterial spindle cell pseudotumor
- Fibromatosis
- Dermatomyofibroma
- Myoid fibroma of the skin
- Cellular schwannoma
- Leiomyoma (pilar, vascular type, ordinary)
- Myoepithelioma (including storiform, herring-bone)
- Dermatofibrosarcoma protuberans
- Dermatofibrosarcoma protuberans with fibrosarcomatous areas (herring-bone)
- Pigmented dermatofibrosarcoma protuberans (Bednar tumor)
- Bednar tumor with fibrosarcomatous areas (herring-bone)
- Fibrosarcoma (herring-bone)
- Leiomyosarcoma
- Myofibrosarcoma
- Spindle cell liposarcoma
- Malignant peripheral nerve sheath tumor
- Fibrous monophasic synovial sarcoma
- Fibrous synovial sarcoma (herring bone)
- Leiomyomatous (spindle cell) rhabdomyosarcoma in children
- Spindle cell rhabdomyosarcoma in adults
- (Extra-)gastrointestinal stromal tumor and other intra-abdominal spindle cell lesions
- Follicular dendritic cell sarcoma
- Cytokeratin-positive fibroblastic reticulum cell neoplasm
- Melanoma

B. Tumors with storiform (cartwheel) pattern
- Benign fibrous histiocytoma
- Neurofibroma
- Myoepithelioma
- Cellular perineurioma (storiform perineurial fibroma)
- Dermatofibrosarcoma protuberans (classic type)
- Pigmented dermatofibrosarcoma protuberans (Bednar tumor)
- Malignant fibrous histiocytoma (storiform-pleomorphic)
- Metastatic sarcomatoid renal cell carcinoma

C. Tumors and pseudotumors with whorled / curlicue pattern
- Pacinian neuroma
- Benign fibrous histiocytoma (epithelioid type with cellular whorls) (occasional)
- Primary cutaneous and metastatic meningioma
- Meningioma-like tumor of the skin
- Nerve sheath tumor (benign, and malignant)
- Nerve sheath myxoma (Pacinian neurofibroma)
- Perineurioma (mainly cellular and Pacinian types)
- Myopericytoma-type perivascular myoma
- Plexiform fibrohistiocytic tumor
- Spindle cell liposarcoma
- Dedifferentiated liposarcoma (with meningothelial whorls)
- Melanoma

D. Tumors and pseudotumors with palisading pattern
- Traumatic neuroma
- Palisading cutaneous fibrous histiocytoma
- Dermatofibroma with myofibroblastic differentiation
- Palisaded angioleiomyoma
- Benign nerve sheath tumor
- Palisaded encapsulated neurona / solitary circumscribed neurona of the skin
- Myoepithelioma
- Dermatofibrosarcoma protuberans (occasional)
Primary cutaneous and metastatic meningioma
- Neuroblastoma-like neurilemoma
- Leiomyoma
- Leiomyosarcoma
- Epithelioid sarcoma
- Fibrous synovial sarcoma (rare)
- (Extra-)gastrointestinal stromal tumor
- Malignant melanoma

E. Tumors with amianthoid structures and (pseudo)rosettes
- Dendritic cell neurofibroma with pseudorosettes
- Myxopapillary ependymoma
- Low-grade fibromyxoid sarcoma
- Hyalinizing spindle cell tumor with giant rosettes
- Intranodal palisaded myofibroblastoma
- Soft tissue myofibroblastoma
- Schwannoma
- Soft tissue perineurioma
- Peripheral neuroepithelioma (PNET) / Ewing's sarcoma
- Neuroblastoma
- Malignant peripheral nerve sheath tumor (rare)
- Malignant lymphoma

F. Tumors with cording / chordoid pattern
- Calcifying aponeurotic fibroma
- Chondroid lipoma
- Myoepithelioma
- Mixed tumor
- Parachordoma
- Epithelioid hemangioendothelioma
- Extraskeletal myxoid chordrosarcoma, including the neuroendocrine variant
- Round cell liposarcoma (rare)
- Epithelioid variant of malignant peripheral nerve sheath tumor
- Metastatic chordoma
- Ossifying fibromyxoid tumor
- Epithelioid sarcoma

G. Tumors with endocrinoid pattern
- Paraganglioma (zellballen)
- Glomus tumor
- Alveolar rhabdomyosarcoma, solid variant
- Paraganglioma-like dermal melanocytic tumors
- Metastatic renal cell carcinoma
- Melanoma

H. Tumors with alveolar pattern
- Alveolar rhabdomyosarcoma
- Alveolar soft part sarcoma
- Ewing's sarcoma (pseudoalveolar)
- Metastatic renal cell carcinoma
- Melanoma

I. Tumors with pseudo-glandular pattern
- Adenomatoid tumor (lace-like or tubular pattern)
- Pseudoglandular schwannoma
- Tenosynovial giant cell tumor – diffuse type (cleft-like “pseudosynovial” spaces)
- Monophasic epithelial synovial sarcoma
- Monophasic epithelial and biphasic mesothelioma
- PNET with adamantinomatoid pattern
- Melanoma
J. Tumors with pseudo-vascular pattern

- Hamartoma of the scalp with pseudoangiosarcomatous features
- Pseudoangiomatous hyperplasia of mammary stroma
- Giant cell fibroblastoma
- Spindle cell lipoma – pseudoangiomatous variant
- Angiomyofibroblastoma
- Epithelioid sarcoma
- Rhabdomyosarcoma – sclerosing, pseudovascular variant
- Melanoma

K. Tumors with retiform pattern

i. Reticular – Netlike
   - Reticular perineurioma
   - Extraneurial retiform perineurioma
   - Hybrid perineurioma-schwannoma and perineurioma-neurofibroma tumors
   - Myoepithelioma

ii. Rete Testis-like
   - Retiform hemangioendothelioma
   - Composite hemangioendothelioma
   - Polymorphous hemangioendothelioma

L. Tumors with (pseudo-)papillary pattern

- Lipoma arborescens
- Spindle cell lipoma (occasional)
- Dabska's tumor
- Tenosynovial giant cell tumor – diffuse type
- Extraspinal ependymoma (myxopapillary type)
- Mesothelioma
- Melanoma
Table VII. Distinctive vascular patterned lesions (mainly non-endothelial).

A. Tumors with constant hemangiopericytoma-like pattern 26 63 64
- Nasopharyngeal angiofibroma
- Myofibroma and myofibromatosis
- Myopericytoma
- Solitary fibrous tumor
- Hemangiopericytoma
- Extraskeletal mesenchymal chondrosarcoma
- Extrarenal renin-producing juxtaglomerular tumor
- Spindle cell synovial sarcoma

B. Tumors with possible hemangiopericytoma-like pattern 26 63 64
- Pyogenic granuloma
- Hemangiopericytoma-like dermatofibroma
- Epithelioid benign fibrous histiocytoma
- Cellular fibrous histiocytoma
- Kaposi's sarcoma
- Infantile fibrosarcoma
- Spindle cell lipoma with pseudoangiomaotus pattern
- Malignant fibrous histiocytoma (storiform-pleomorphic)
- Malignant peripheral nerve sheath tumor
- Low grade fibromyxoid sarcoma (sometimes in recurrence)
- Synovial sarcoma
- Phosphaturic mesenchymal tumor
- Leiomyosarcoma
- Liposarcoma (pleomorphic; dedifferentiated)
- Glomus tumor (glomangiopericytoma)
- Paraganglioma
- Thymoma (predominantly spindle cell type / medullary type / type A)
- Low grade endometrial stromal sarcoma - primary retroperitoneal; metastatic
- Malignant mesothelioma – fibrous type
- Metastatic sarcomatoid carcinoma (kidney, lung)
- Melanoma – primary, metastatic 59

C. Tumors with prominent perivascular hyalinization
- Orbital & extraorbital giant cell angiofibroma
- Schwannoma
- Cellular angiofibroma (external genital sites)
- Aggressive angiomyxoma
- Solitary fibrous tumor
- Pleomorphic hyalinizing angiectatic tumor
- Hemangiopericytoma
- Extragastrointestinal gastrointestinal stromal tumor (not uncommon)

D. Tumors with plexiform capillary pattern
- Paraganglioma (mainly “branchiomeritic type”)
- Hemangioblastoma, extra-neuraxial
- Myxoid liposarcoma 65
- Myxoid malignant fibrous histiocytoma

E. Tumors with glomeruloid capillary pattern
- Intravascular papillary endothelial hyperplasia - Masson’s phenomenon
- Tufted angioma
- Glomeruloid hemangioma
- Dabska’s tumor 66
- Malignant angioendetheliomatosis (intravascular lymphoma)
- Some neural and neuroendocrine neoplasms
F. Lesions with hemorrhagic changes / pattern

- Organizing hematoma
- Pyogenic granuloma
- Kaposi’s sarcoma
- Soft tissue Kaposiform hemangioendothelioma
- Capillary hemangioma of the infantile hemangioendothelioma type
- Hemorrhagic dermatomyofibroma
- Intranodal hemorrhagic spindle cell tumor with “amianthoid” fibres
- Extraskeletal myxoid chondrosarcoma
- Pleomorphic hyalinizing angiectatic tumor

G. Tumors with aneurysmal changes / pattern

- Cavernous hemangioma, sinusoidal hemangioma, ...
- Angiomatoid cellular blue nevus
- Aneurysmal bone cyst of soft tissue
- Aneurysmal (“angiomatoid”) benign fibrous histiocytoma
- Angiomatoid fibrous histiocytoma (angiomatoid “malignant fibrous histiocytoma”)
- Atypical fibroxanthoma (aneurysmal or pigmented variant)
- Dermatofibrosarcoma protuberans
- Malignant fibrous histiocytoma giant-cell type
- Soft tissue giant cell tumor
- Pleomorphic hyalinizing angiectatic tumor
- Cutaneous Ewing’s sarcoma
- Paraganglioma (occasional)
- Soft tissue telangiectatic osteosarcoma
- Metastatic sarcomatoid carcinoma (mainly from kidney)
- High grade sarcoma of various types (e.g. angiomatoid or angiectoid or angiosarcoma-like variant of epithelioid sarcoma)
Table VIII. Combined, polyphasic, and heterologous tumors (other than collision tumors).

A. Combined (homologous and heterologous) tumors
- Combined dermatofibroma (two variants in the same lesion)
- Giant cell fibroblastoma / dermatofibrosarcoma protuberans
- Giant cell fibroblastoma / Bednar tumor
- Giant cell angiofibroma / dermatofibrosarcoma protuberans
- Bednar tumor / dermal melanocytosis
- Dermatofibrosarcoma protuberans / myxofibrosarcoma
- Lipoma / leiomyoma
- Angiomyomatosis / glomus cell proliferation
- Acquired tufted angioma / vascular malformation
- Lipoleiomyosarcoma
- Neurofibroma / schwannoma
- Neurofibroma / perineurioma 68
- Schwannoma / perineurioma 69
- Sarcomas (usually angiosarcomas) in malignant peripheral nerve sheath tumor
- Mesenchymoma
- Soft tissue ectomesenchymoma (benign and malignant Triton tumor, ...)
- Angiosarcoma in neurofibroma
- Epithelioid malignancies in schwannomas
- Melanoma with adenoid pattern 59, 59a
- Melanoma with neurosarcomatous transformation
- Melanoma with ganglioneuroblastic differentiation
- Rhabdomyosarcoma in a congenital melanocytic nevus

B. Biphasic homologous (bimorphic) tumors
- Lymphangiomyoma
- Schwannoma (Antoni A & B areas).
- Myofibroma and myofibromatosis
- Fibrous hamartoma of infancy
- Myolipoma (lipoleiomyoma)
- Lipofibromatosis
- Angiolipoma
- Cellular angiolipoma
- Angioleiomyoma
- Neural fibrolipoma (fibrolipomatous hamartoma of nerve)
- Lipomatous neurofibroma
- Myelolipoma (adrenal; extraadrenal)
- Fibrohistiocytic lipoma
- Kaposi’s sarcoma
- Plexiform fibrohistiocytic tumor
- Dermatofibrosarcoma protuberans (with myoid differentiation; with nodular sclerotic changes)
- Superficial / cutaneous angiomyxoma with epithelial component
- Low grade fibromyxoid sarcoma
- Hemosiderotic fibrohistiocytic lipomatous tumor
- Intramuscular hemangioma with fatty component (can be viewed as triphasic)
- Spindle cell lipoma in intramuscular lipoma (can be viewed as triphasic) 70
- Angiomyolipoma (can be viewed as triphasic)
- Synovial sarcoma
- Mixed-type liposarcoma (dedifferentiated liposarcoma) 71
- Low-grade dedifferentiated liposarcoma 72
- Mixed-type liposarcoma (combined type: myxoid/round cell and pleomorphic liposarcoma)
- Lipoleiomyosarcoma
- Myxoid liposarcoma with nodules of round cell liposarcoma
- Lipomatous hemangioepicyctoma
- Extraskeletal mesenchymal chondrosarcoma
- Dedifferentiated parosteal osteosarcoma
- Ganglioneuroma (mature Schwannian stroma and ganglion cells)
- Ganglioneuroblastoma
- Desmoplastic small round cell tumor (nests of undifferentiated tumor cells plus fibrous stroma)
- Merkel cell tumor with squamous cell differentiation, and/or glandular differentiation

C. Biphasic heterologous tumors – with epithelial elements
- Hyaline cell rich-chondroid syringoma
- Mixed tumors
- Myoepitheliomas
- Parachordoma
- Lipoma with synovial metaplasia
- Ectopic hamartomatous thymoma
- Superficial angiomyxomas / cutaneous myxomas
- Schwannoma (with glandular differentiation; with squamous cell elements)
- Malignant peripheral nerve sheath tumor with glandular and neuroendocrine differentiation; with squamous cell islands
- Ectomesenchymoma (benign, and malignant)
- Malignant glandular Triton tumor
- Biphasic synovial sarcoma with glandular differentiation
- Epithelial synovial sarcoma with squamous cell differentiation
- Pretilial soft tissue adamantinoma
- Merkel cell tumor with leiomyo- or rhabdomyosarcomatous differentiation (see below at “E”)
- Dermal squamomelanocytic tumor
- Dermal basomelanocytic tumor

D. Pseudo-biphasic tumors
- Tumor growing in normal tissue
  i. Intramuscular lipoma
  ii. Intramuscular hemangioma
  iii. Intramuscular fasciitis
  iv. Proliferative myositis
  v. Fibromatosis coli
  vi. Intraneural leiomyoma
  vii. Intraneural glomus tumor
  viii. Intraneural synovial sarcoma
  ix. Sarcomas infiltrating fat
- Tumor growing in tumor
  i. Schwannoma with metastatic carcinoma of the breast
  ii. Meningioma with metastatic carcinoma (various sources)
  iii. Malignant transformation in a benign tumor (e.g. angiosarcoma in hemangioma, Dabska tumor in a lymphangiomia, malignant peripheral nerve sheath tumor in a ganglioneuroma, carcinoma arising in ectopic hamartomatous thymoma, carcinoma arising in a mixed tumor of soft tissue, …)

E. Biphasic tumors with skeletal muscle differentiation
- Rhabdomyomatous mesenchymal hamartoma of the skin
- Leiomyoma and leiomyosarcoma
- Dedifferentiated liposarcoma
- Myxoid liposarcoma
- Dedifferentiated chondrosarcoma
- Neuromuscular choristoma (benign Triton tumor / benign ectomesenchymoma)
- Malignant peripheral nerve sheath tumor / Malignant ectomesenchymoma (malignant Triton tumor)
- Benign and malignant mesenchymoma
- Ganglioneuroma and ganglioneuroblastoma
- Merkel cell tumor
- Metastasis from any tumors of any lineage other than mesenchymal, possibly containing skeletal muscle component: germ cell tumors, blastomas, carcinomas, tumors of specialized stroma (phyllodes tumors of breast, uterine adenosarcoma, Sertoli-Leydig cell tumor of the ovary), neuroendocrine carcinomas (Merkel cell tumor and other small cell carcinomas)
Table IX. Histogenetically diverse but morphologically similar tumors and pseudotumors with ...

A. Round-cell / epithelioid / plasmacytoid features
- Epithelioid benign fibrous histiocytoma
- Myoepithelioma
- Mixed tumor
- Hyaline cell rich-chondroid syringoma
- Parachordoma
- Epithelioid neurofibroma
- Epithelioid schwannoma
- Cutaneous epithelioid Schwannoma
- Epithelioid hemangioma
- Glioma tumor
- Epithelioid leiomyoma
- Epithelioid angioleiomyoma
- Tenosynovial giant cell tumor – diffuse type
- Angiomyofibroblastoma
- Solitary fibrous tumor
- Ossifying fibromyxoid tumor
- Epithelioid hemangioendothelioma
- Epithelioid sarcoma
- Epithelioid sarcoma-like hemangioendothelioma
- Epithelioid leiomyosarcoma
- Epithelioid variant of malignant peripheral nerve sheath tumor
- Epithelioid angioleiomyoma
- Epithelioid angiomyolipoma and other PEComas
- Epithelioid angiosarcoma
- Poorly differentiated fibrosarcoma
- Sclerosing epithelioid fibrosarcoma
- Round cell liposarcoma
- Epithelioid liposarcoma
- Extraskelatal myxoid chondrosarcoma – epithelioid variant
- Alveolar soft part sarcoma
- Proximal-type epithelioid sarcoma
- Extranreal malignant rhabdoid tumor
- Biphasic & monophasic epithelial synovial sarcoma
- Biphasic & monophasic epithelial mesothelioma
- Deciduoid mesothelioma
- Extramedulamp epithelioid histiocytic sarcoma

B. Clear cell changes
- Fibrous papule of the nose, clear cell variant
- Clear cell dermatofibroma
- Myoepithelioma – clear cell type
- Angioleiomyoma – clear cell variant
- Angiomyolipoma
- Leiomyoma – clear cell variant
- Atypical fibroxanthoma – clear cell type
- PEComas of soft tissue
- PEComas of the skin
- Distinctive dermal clear cell mesenchymal neoplasm
- Epithelioid leiomyosarcoma
- Epithelioid malignant peripheral nerve sheath tumor
- Sclerosing epithelioid fibrosarcoma
- Alveolar rhabdomyosarcoma – clear cell variant
- Ewing’s sarcoma – clear cell variant
- Clear cell sarcoma
- Ectopic hamartomatous thymoma with cords of clear cells
- Alveolar soft part sarcoma (clear cell degenerative variant)
- Balloon cell nevus
- Balloon cell blue nevus
- Balloon cell melanoma
- Paraganglioma-like dermal melanocytic tumor
- Metastasis of clear cell tumors from other organs
- Peripheral T-cell lymphoma “unspecified”
C. (Uni-/multi-)vacuolated (pseudo-)lipoblasts, and (pseudo-)physalipherous cells

- Silicone granuloma
- Fat necrosis
- Atrophic fat in malnutrition
- Polyvinylpyrrolidone pseudotumoral histiocytic reaction (“mucicarminophilic histiocytosis”)
- Hibernoma
- Rhabdomyoma (secondary vacuoles to loss of glycogen)
- Chondroid lipoma
- Pseudolipoma (battered buttock syndrome)
- Spindle cell hemangioma
- Parachordoma
- Paraganglioma
- Adenomatoid tumor (pseudovacuolated cells)
- Epithelioid hemangioendothelioma
- Inflammatory myxohyaline tumor
- Low grade myxofibrosarcoma (myxoid fibrosarcoma or myxoid malignant fibrous histiocytoma gr. I)
- Myxoid malignant fibrous histiocytoma (gr. II)
- Liposarcoma
- Pseudolipoblastic (“signet-ring”) nerve sheath tumor
- Lipoblastic meningioma
- Chordoma - metastatic, primary heterotopic, periphericum
- Ewing’s sarcoma
- (Extra-)gastrointestinal stromal tumor
- Large cell lymphoma (signet ring cell type)
- Melanoma (signet ring-cell type - primary, and metastatic)
- Metastatic adenocarcinoma

D. Eosinophilic / oncocytic features

- Silica reaction
- Crystal storing histiocytosis
- Rhabdomyoma – oncocytic type
- Glomus tumor – oncocytic type
- Paraganglioma – oncocytic type
- Oncocytic myoepithelioma
- Soft tissue oncocytoma
- Extraskeletal myxoid chondrosarcoma (cellular variant)
- Alveolar soft part sarcoma
- Malignant extrarenal rhabdoid tumor
- Proximal-type epithelioid sarcoma
- Some benign and malignant melanocytic tumors

E. Granular cells

- Granular fibrous papule of the face
- Granular histiocytic cell reaction
- Granular stromal cell reaction with various tumors
- Granular cell tumor / granular cell schwannoma
- Congenital gingival granular cell tumor
- Granular cell dermatofibroma
- Atypical fibroxanthoma with granular cells
- Dermatofibrosarcoma protuberans with granular cells
- Primitive polypoid granular cell tumor (of the skin)
- Primitive nonneural granular cell tumors of skin (the same as the previous one)
- Granular cell perineurioma
- Granular cell leiomyoma
- Hibernoma
- Malignant granular cell tumor
- Granular cell leiomyosarcoma
- Granular cell angiosarcoma
- Some benign and malignant melanocytic tumors

F. Rhabdoid- / pseudo-rhabdoid features

- Crystal-storing histiocytosis
- Adult rhabdomyoma
- Rhabdomyomatous mesenchymal hamartoma
- Myoepithelioma
<table>
<thead>
<tr>
<th>Lesions of Soft Tissue</th>
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<tbody>
<tr>
<td>Leiomyosarcoma with rhabdoid features</td>
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<tr>
<td>Rhabdomyosarcoma with rhabdoid features</td>
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<tr>
<td>Synovial sarcoma</td>
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<tr>
<td>Extraskeletal myxoid chondrosarcoma – rhabdoid variant</td>
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<tr>
<td>Epithelioid malignant peripheral nerve sheath tumor</td>
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<tr>
<td>Proximal-type epithelioid sarcoma</td>
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<tr>
<td>Extraskeletal malignant peripheral nerve sheath tumor</td>
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<tr>
<td>Desmoplastic small round cell tumor</td>
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<td>Mesothelioma</td>
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<tr>
<td>Extraskeletal myxoid chondrosarcoma – rhabdoid variant</td>
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<tr>
<td>Embryonal rhabdomyosarcoma</td>
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<td>Neuroblastoma</td>
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<td>Desmoplastic small round cell tumor</td>
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<tr>
<td>Extraskeletal mesenchymal chondrosarcoma</td>
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<tr>
<td>Extraskeletal myxoid chondrosarcoma – cellular variant</td>
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<tr>
<td>Small cell osteosarcoma</td>
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<tr>
<td>Malignant melanoma</td>
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<tr>
<td>Small cell carcinoma (primary: Merkel cell-type, and non-Merkel cell type; metastatic: pulmonary, and extrapulmonary)</td>
</tr>
</tbody>
</table>

G. Ganglion / ganglion-like cells

- Ischemic fasciitis (Atypical decubital fibroplasia)
- Ganglion cell choristoma of skin
- Cutaneous ganglion cell tumor
- Cutaneous ganglioneuroma
- Desmoplastic cutaneous ganglioneuroma
- Proliferative fasciitis
- Proliferative myositis
- Ganglioneuroma

H. Small round cell tumors

- Glomus tumors (benign, borderline, malignant)
- Tenosynovial giant cell tumor
- Primitive neuroectodermal tumor / extraskeletal Ewing’s sarcoma
- Malignant hemangiopericytoma
- Round cell liposarcoma
- Embryonal rhabdomyosarcoma
- Neuroblastoma
- Desmoplastic small round cell tumor
- Extraskeletal malignant peripheral nerve sheath tumor
- Embryonal and alveolar rhabdomyosarcoma
- Extraskeletal mesenchymal chondrosarcoma
- Extraskeletal myxoid chondrosarcoma – cellular variant
- Small cell osteosarcoma
- Malignant lymphoma
- Granulocytic sarcoma / leukaemia
- Extramedullary myeloid cell tumors
- Malignant melanoma
- Small cell carcinoma (primary: Merkel cell-type, and non-Merkel cell type; metastatic: pulmonary, and extrapulmonary) |

I. Pleomorphic soft tissue tumors

- Benign or intermediate
  i. Pleomorphic fibrous papule of the face
  ii. Pleomorphic fibroma of the skin
  iii. Subungal pleomorphic fibroma
  iv. Subungal myxoid pleomorphic fibroma
  v. Pleomorphic hyalinizing angiectatic tumor
  vi. Hemosiderotic fibrohistiocytic lipomatous tumor
  vii. Atypical fibroxanthoma
- Malignant
  viii. Storiform-pleomorphic malignant fibrous histiocytoma
  ix. Dedifferentiated areas in well-differentiated liposarcoma
  x. Pleomorphic liposarcoma
  xi. Pleomorphic leiomyosarcoma
  xii. Pleomorphic myofibrosarcoma
  xiii. Pleomorphic rhabdomyosarcoma
  xiv. Pleomorphic malignant peripheral nerve sheath tumor
  xv. Pleomorphic intimal sarcoma
  xvi. Malignant PEC-oma (may mimic pleomorphic myogenic sarcoma)
Table X. Tumors and pseudotumors with diagnostic clues.

A. Architecturally distinct features

- Swiss cheese pattern (oleogranuloma / paraffinoma, silicone granuloma, fat necrosis, chalazion, …)
- Verocay bodies and Verocay-like bodies or nuclear palisading (benign and malignant nerve sheath tumors, leiomyoma, leiomyosarcoma, spindle cell synovial sarcoma, extragastrointestinal gastrointestinal stromal tumors)
- Homer-Wright rosettes (neuroblastoma, Ewing’s sarcoma, PNET, Merkel cell carcinoma) and perivascular pseudorosettes (ependymoma)
- Amianthoid fibers, or giant collagen rosettes (palisaded myofibroblastoma, hyalinizing spindle cell tumor with giant rosettes, dendritic cell neurofibroma with pseudorosettes and other nerve sheath tumors, solitary fibrous tumor, osteocartilaginous tumors, others)
- Nonspecific rosettes (non-Hodgkin lymphoma – occasional; epithelioid osteosarcoma, melanoma)
- “Zellballen” (paraganglioma)
- Cambium layer (botryoid embryonal rhabdomyosarcoma)
- Filigree pattern (Ewing’s sarcoma)
- Light cell/dark cell phenomenon – marble appearance (Ewing’s sarcoma, MPNST)
- Azzopardi’s effect (Merkel cell tumor; other small cell tumors)
- Zoning phenomenon from fibroblastic center to ossified periphery (myositis ossificans)
- Reverse zoning phenomenon (osteosarcoma)
- Pacinian corpuscle (Pacinian neuroma)
- Wagner-Meissner body (diffuse neurofibroma)
- Pacinian body-like and Meissner body-like structures (nerve sheath tumors)
- Onion-bulb structures (hereditary diffuse hypertrophic neuropathies: Charcot-Marie-Tooth disease, Dejerine-Sottas disease; other true onion bulb neuropathies: Refsum disease, Krabbe’s disease, Roussy-Levi disease, metachromatic leukodystrophy, neurofibromatosis type 1, diabetic neuropathy, chronic inflammatory demyelinating polyradiculoneuropathy; localized hypertrophic inflammatory neuropathy; “localized hypertrophic neuropathy” as true onion bulb neuropathy)
- Pseudo-onion bulb structures (“localized hypertrophic neuropathy” as intraneural perineurioma; extraneural/soft tissue perineurioma)
- Cytophagocytosis (cytophagic histiocytic panniculitis/subcutaneous panniculitis-like T-cell lymphoma; inflammatory myxohyaline tumor)
- Petaloid globules – chenille bodies (elastofibroma)
- Skeinoid fibres (intestine-based stromal tumor with neural differentiation, and rarely extra-gastrointestinal GIST)
- Ferruginous bodies – asbestos bodies (mesothelioma)
- Intravascular fibrin thrombi (-angiolipoma)

B. Extracellular findings / stromal precipitates

- Schaumann bodies (sarcoid, sarcoid-like granulomatous reactions)
- (Pseudo-)psammomatous calcifications (calcifying fibrous pseudotumor)
- Psammoma bodies (cutaneous meningioma)
- Congophilic green birefringent material (amyloid tumor)
- Non-congophilic amorphous material in pelvic and retroperitoneal soft tissue (Tamm-Horsfall protein deposition)
- Non-congophilic positively birefringent microcrystals (tumoral pseudogout)
- Non-congophilic, needle-shaped birefringent crystals (tophaceous gout)
- Foreign bodies (even partly intracellular) (silica [soil, glass], talc, starch, condom emulsion, silicone, cactus spine, sea-urchin spine, hairs, wooden splinters, suture material, dacron graft, woollen swab, …) (foreign body reactions and granulomas, pseudotumoral granulomatous reactions, textilomas, …)
- Non-congophilic amorphous material in pelvic and retroperitoneal soft tissue (Tamm-Horsfall protein deposition)
- Petaloid globules – chenille bodies (elastofibroma)
- Skeinoid fibres (intestine-based stromal tumor with neural differentiation, and rarely extra-gastrointestinal GIST)
- Ferruginous bodies – asbestos bodies (mesothelioma)
- Intravascular fibrin thrombi (-angiolipoma)

C. Cytohistologically distinct features

- Cell peculiarities
  - Fleurette-like cells (pleomorphic lipoma, giant cell angiofibroma)
  - Toutou cells (juvenile xanthogranuloma, Erdheim-Chester disease)
  - Reed-Sternberg-like cells (inflammatory myxoid-hyaline tumor, proliferative myosis, some extranodal soft tissue large cell lymphomas, …)
  - Spider cells (rhabdomyoma)
  - Tombstone-cells (epithelioid hemangioma)
  - Hobnail-cells (hob-nail hemangiomas – Dabska’s type, retiform, polymorphous hemangioendothelioma, …)
  - Vacuolated cells (lipoblast → liposarcoma; lipoblast-like stromal cells → myxofibrosarcoma; lipoblast-like macrophage → fat necrosis, oleogranulomas; bubbly histocytes → polyvinilpyrrolidone reaction; endothelioblast and endotheliocyte → vascular tumors, …)
Strap cells / cross-striated cells (rhabdo-myomatous/myosarcomatous tumors)
- Longitudinally pseudostriated cells (crystal storing histiocytosis in lymphoproliferative disorders)

**Intracellular findings**
- Melanin (melanocytic tumor, neural tumor, neuroectodermal tumor)
- Intracytoplasmic hyaline globules (Kaposi's sarcoma, glomeruloid hemangioma, Kaposiform hemangioendothelioma)
- Trichrome stain positive intracytoplasmic inclusions (infantile digital fibromatosis, myofibrosarcoma)
- Granular material in reactive histiocytes (site of previous surgery)
- Intracytoplasmic PAS-D resistant bodies (granular cell tumor, extraintestinal Whipple's disease)
- Intracellular crystalline PAS-D positive material (alveolar soft part sarcoma)
- Intracytoplasmic paracrystalline material (crystal storing histiocytosis in lymphoproliferative disorders)
- Michaelis-Gutmann bodies (malakoplakia)
- Asteroid bodies (sarcoïd, sarcoïd-like granulomatous reactions, ...)
- Infectious bacilli (bacillary angiomatosis, mycobacterial disease, atypical mycobacterial disease, Whipple disease)
- Cytoplasmic cross-striations (rhabdo-myomatous / -myosarcomatous tumors)
- Cytoplasmic fibrillary pattern (“wrinkled cigarette paper” in Gaucher disease, ...)
- Cytoplasmic intranuclear pseudoinclusions (meningioma, nerve sheath tumors, melanocytic tumors, soft tissue lymphoplasmacytic proliferative disorders [Dutch bodies], metastatic hepatocellular carcinoma)
- Nuclear vacuoles – lochkern (normal fat)
- Nuclear grooves (soft tissue metastatic thyroid papillary carcinoma; metastatic granulosa cell tumor; melanoma)
Table XI. Histologically* pigmented tumors and pseudotumors with ...

A. Melanin pigment

- Fibrous papule of face —pigmented variant
- Cutaneous benign fibrous histiocytoma (clinically pigmented)
- Intradermal Spitz nevus
- Dermal dendritic melanocytic proliferations (including deep seated blue nevus)
- Pigmented pilomatrixoma and melanocytic pilomatrixoma
- Pigmented (melanotic) neurofibroma
- Pigmented (melanotic) schwannoma
- Psammomatous melanotic schwannoma
- Cutaneous psammomatous melanotic schwannoma
- Pigmented dermatofibrosarcoma protuberans (Bednar tumor)
- Bednar tumor associated with dermal melanocytosis
- Neurocristic cutaneous hamartoma
- Neurocutaneous melanosis (melanoma, melanoblastoma, meningeal melanocytosis)
- Pigmented (melanotic) neuroectodermal tumor of infancy
- Ordinary melanoma
- Primary dermal melanoma
- Desmoplastic melanoma
- Deep melanoma
- Metastatic melanoma
- Pigmented epithelioid melanocytoma / animal-equine-type melanoma / epithelioid blue nevus (sporadic, and syndromic – associated with Carney’s syndrome)
- Clear cell sarcoma
- Pigmented ganglioneuroblastoma
- Phaeochromocytoma and other extraadrenal paraganglioma
- Deep melanoma

B. Hemosiderin

- Hemorrhage / hematoma / ancient hematoma
- Hemangiomas (mainly cavernous type) and phakomatoses
- Synovial hemangioma
- Hemosiderotic dermatofibroma
- Targetoid hemosiderotic hemangioma
- Aneurysmal benign fibrous histiocytoma
- Aneurysmal (pigmented) atypical fibroxanthoma
- Kaposi’s sarcoma
- Pleomorphic hyalinizing angiectatic tumor
- Hemosiderotic fibrohistiocytic lipomatous tumor
- Diffuse-type giant cell tumor (pigmented villonodular synovitis / pigmented villonodular bursitis)
- Angiomatoid fibrous histiocytoma
- Tenosynovial giant cell tumor (localized and diffuse type)
- Clear cell sarcoma

C. Purified carbon / charcoal

- Preoperative localization of gut biopsy sites
- Preoperative localization of nonpalpable tumor lesions

D. India Ink (rare), Cinnabar, Chrome green, Cobalt blue

- Delayed hypersensitivity reactions to tattoos

E. Heavy metals particles

- Paraarticular histiocytic proliferation induced by cobalt-chromium and titanium with intracytoplasmic small black particles in histiocytes (prosthetic heavy metal detritus)

* Clinically pigmented or erythematous tumors and pseudotumors without pigment at histology: Aschoff’s nodules (rheumatic fever), Farber’s lipogranulomatosis, Sweet’s syndrome, panniculitis, juvenile xanthogranuloma, mastocytoma, cutaneous T-cell lymphoma, metastatic cutaneous neuroblastoma
Table XII. Clinically alarming benign tumors and pseudotumors.

A. Multiple / multicentric, possibly recurring benign lesions

- Cutaneous sarcoid
- Papular angiookeratoma (a frequent mimicker of melanoma)
- Multiple eruptive angiofibromas
- Multiple eruptive dermatofibromas (in HIV and immunosuppressed patients)
- Recurrent pyogenic granuloma with satellitosis
- Multiple pyogenic granulomas
- Generalized pyogenic granuloma
- Multiple nevus lipomatosus cutaneous superficialis
- Multiple cutaneous pilar leiomyomas
- Multiple cutaneous chondromas
- Multiple milary osteomas of the face
- Nodular fasciitis (satellite nodules)
- Infantile fibromatosis (desmoid-type)
- Juvenile hyaline fibromatosis
- Juvenile lipofibromatosis
- Infantile digital fibromatosis - recurring
- Multiple juvenile xanthogranuloma
- Multicentric reticulohistiocytosis / multiple reticulohistiocytic granulomata / progressive nodular histiocytoma / generalized eruptive histiocytoma, xanthoma disseminatum
- Tendinous xanthomas
- Xanthogranulomatous inflammation (e.g. in Erdheim-Chester disease)
- (Recurring) tumoral calcinosis
- (Recurring) tophaceous pseudogout (CPPD crystal deposition disease - tumoral form)
- Infantile myofibromatosis
- Multiple multifocal (adult) rhabdomyomas of the neck
- Multiple intramuscular myxomas
- Myolipoma (of soft tissue)
- Inflammatory pseudotumor / inflammatory myofibroblastic tumor
- Infantile hemangiopericytoma
- Multiple glomus tumors
- Glomangiomatosis (may recur)
- Multiple glomangiomyomas
- Infiltrating (aggressive, atypical) glomus tumor
- Recurring glomus tumor
- Plexiform neurofibroma
- Multiple schwannomas (schwannomatosis)
- Multiple cutaneous plexiform schwannomas
- Multiple melanotic schwannomas
- Multiple cellular neurothekeomas
- Giant cell fibroblastoma (recurring)
- Multiple lipomas
- Multiple spindle cell lipomas (familial and non-familial forms)
- Multiple angiolipomas (familial and non-familial forms)
- Symmetric lipomatosis
- Diffuse lipomatosis
- Diffuse angiomatosis
- Multiple spindle cell hemangi(endothelio)mas
- Spindle cell hemangiomatosis
- Fibrodyplasia (myositis) ossificans progressiva
- (Recurring) bizarre parosteal osteochondromatous proliferation (Nora’s disease)
- Multiple (multifocal) leiomyomas
- Leiomyomatosis peritonealis disseminata
- Granular cell tumor
- Progressive lymphangioma (benign lymphangioendothelioma)
- Reactive angioendotheliomatosis
- (Epithelioid hemangioendothelioma)
- Kaposi’s sarcoma
- Cerebriform fibrous proliferations in Proteus syndrome
• (Some) multiple smooth muscle tumors in immunocompromised patients
• Paraganglioma (multicentric)
• Reactive nodular fibrous pseudotumors (of the gastrointestinal tract and mesentery)
• Sclerosing mesenteritis, mesenteric panniculitis, mesenteric lipodystrophy, and other inflammatory pseudotumors
• Multiple fibrosclerosis (any combination of Ormond’s disease, sclerosing cholangitis, mediastinal fibrosis, Riedel’s thyroiditis, pseudotumor of the orbit, autoimmune pancreatitis)
• Sclerosing mediastinitis (idiopathic fibroinflammatory lesions of the mediastinum)

B. Painful soft tissue tumors and pseudotumors
• Traumatic neuroma
• Keloid (sometimes)
• Dercum’s disease (adiposis dolorosa)
• Glomus tumors (see section A, above)
• Angiolipoma
• Angioleiomyoma
• Cutaneous leiomyoma
• Morton’s interdigital neuroma
• Schwannomatosis
Table XIII. Pathologically alarming benign tumors and pseudotumors.

A. Lesions with multilobular / mosaic growth pattern
- Tumoral calcinosis
- Lipoblastoma
- Cutaneous myxoma
- Neurothekeoma
- Juvenile hemangioma
- Pyogenic granuloma
- Acquired tufted angioma
- Schwannoma
- Extraskeletal chondroma
- Synovial chondromatosis (articular and extra-articular variant)
- Giant cell tumor
- Intramuscular myxoma (occasional)
- Ossifying fibromyxoid tumor (not including here the atypical & malignant variant)

B. Lesions with plexiform / infiltrating margins
- Proliferative fasciitis
- Cranial fasciitis
- Proliferative myositis
- Fibrous hamartoma of infancy
- Fibromatosis (any type, mainly infantile type)
- Plexiform xanthomatous tumor
- Plexiform Schwannoma (cutaneous and subcutaneous, and deep forms)¹⁰⁸a, Schwannomatosis ¹⁰⁸b-d
- Plexiform neurofibroma
- Neurothekeoma (classic and cellular type)
- Nerve sheath myxoma ("plexiform myxoma")
- Granular cell tumor (plexiform variant)
- Dendritic fibromyxolipoma
- Intramuscular lipoma (infiltrating lipoma)
- Diffuse lipoblastomatosis (infiltrating lipoblastoma)
- Lipoma arborescens
- Cellular schwannoma (occasional)
- Congenital and childhood plexiform (multinodular) cellular schwannoma ¹¹⁰
- Deep-seated plexiform schwannoma ¹¹¹
- Infiltrating retiform perineurioma ¹¹²
- Plexiform fibrohistiocytic tumor

C. Lesions with intravascular growth
- Intralymphatic granulomas (in Crohn's disease, granulomatous lymphangitis of the scrotum and penis, granulomatous cheilitis, Melkerson and Rosenthal syndrome)
- Organizing thrombus
- Pyogenic granuloma – intravascular variant
- Neurothekeoma
- Epithelioid hemangioma
- Plasma cell granuloma
- Giant cell fibroblastoma
- Intravenous atypical vascular proliferation
- Intravascular papillary endothelial hyperplasia
- Spindle cell hemangioma
- Epithelioid hemangioma
- Myopericytoma – intravascular variant
- Myofibroma (solitary type – adult form)
- Infantile myofibromatosis
- Infantile hemangiopericytoma
- Intravascular glomus tumor
- Intravascular fasciitis
- Angioleiomyoma – intravascular form
- Intravenous leiomyomatosis (conventional and unusual variants) ¹¹⁵
- Myointimoma
- Tenosynovial giant cell tumor
D. Benign lesions with “hypercellularity”

- Cellular fibrous papule of the face
- Cellular digital fibromas
- Cellular dermatofibroma
- Cellular benign fibrous histiocytoma
- Cellular pseudosarcomatous fibroepithelial stromal polyp
- Cellular infantile capillary hemangioma
- Cellular spindle cell hemangioidoendothelioma
- Cellular angiolipoma
- Cellular schwannoma
- Cellular schwannoma with granular cells
- Congenital and childhood plexiform (multinodular) cellular schwannoma
- Cellular neurofibroma
- Cellular leiomyoma
- Cellular perineurioma
- Cellular neurothekeoma
- Infantile fibromatosis (desmoid-type)
- Cellular fibromatosis (palmar type)
- Monophasic cellular variants of myofibromatosis
- Cellular giant cell fibroblastoma
- Cellular intramuscular myxoma
- Cellular rhabdomyoma of fetal type (intermediate variant)
- Cellular tenosynovial giant cell tumor
- Cellular ossifying fibromyxoid tumor
- Cellular blue nevus
Table XIV. Histologically benign lesions (other than vascular proliferations) mistaken for malignant.

A. Tumor-like lesions

- Subcutaneous granuloma annulare (mimicking epithelioid sarcoma, epithelioid angiosarcoma)
- Mitotic granuloma annulare
- Necrobiotic xanthogranuloma
- Malakoplasia
- Nodular fasciitis and variants (intravascular, cranial, peri- and parosteal, intraarticular fasciitis)
- Ischemic fasciitis
- Proliferative fasciitis
- Proliferative myositis
- Desmoid fibromatosis
- Reticulohistiocytoma
- Uterine xanthogranuloma with inconspicuous foam cells and giant cells (mimicking melanoma)
- Polyvinylpyrrolidone pseudotumoral histiocytic reaction ("mucicarminophilic histiocytosis") simulating signet-ring cell carcinoma, and myxoid liposarcoma
- Tumefactive fibroinflammatory lesion
- Xanthogranulomatous pseudotumor (several etiologies, mainly infectious)
- Epithelial sheath neuroma
- Reactive angiendoteliomatosis
- Intramuscular hypertrophic scar following sarcoma resection
- Massive localized lymphedema in morbid obesity
- Pseudoneoplastic skeletal muscle regeneration
- Post-implant pseudoneoplastic parathyromatosis
- Hyperplastic callus in pathologic conditions (e.g. osteogenesis imperfecta)
- Tumefactive soft tissue extension of Paget’s pseudosarcoma
- Tophaceous pseudogout
- Myositis-, panniculitis-, fascitis- and periostitis ossificans
- Fibrodyplasia ossificans progressiva
- Fibroosseous pseudotumor of the digits
- Bizarre parosteal osteochondromatous proliferation (Nora’s lesion)
- Vulvar hypertrophy with lymphedema
- Cutaneous pseudosarcomatous polyp
- Vaginal pseudosarcomatous fibroepithelial polyp
- Pseudotumoral decidualis
- Extramedullary hematopoiesis (e.g. in breast, in retroorbital site, ...)
- Sclerosing extramedullary hematopoietic tumor

B. Tumoral lesions

- Granular cell dermatofibroma
- Benign cellular fibrous histiocytoma
- Atypical (pseudosarcomatous) fibrous histiocytoma of the skin
- Atypical cellular neurothekeoma
- Deeply located benign fibrous histiocytoma
- Intraneural glomus tumor
- Symplastic glomus tumor
- Intraneural leiomyoma
- Granular cell tumor
- Symplastic leiomyoma
- Ancient schwannoma
- Plexiform (multinodular) cellular schwannoma
- Intramuscular hemangioma
- Synovial chondromatosis (extra-articular variant)
- Aggressive angiomyxoma
### Table XV. Reactive endothelial lesions that may histologically mimick malignancy.

- Organizing hematoma
- Ancient hematoma
- Intravascular papillary endothelial hyperplasia
- Acroangiodermatitis
- Glomeruloid hemangioma
- Bacillary angiomatosis
- Acquired progressive lymphangioma / benign lymphangioendothelioma
- Reactive angioendotheliomatosis
- Benign atypical vascular lesions of the lip
- Multiple trauma-induced targetoid hemosiderotic hemangioma-like lesions
- Diffuse dermal angiomatosis of the breast (response to isotretinoin therapy)
- Acquired vulvar lymphangioma (secondary to Crohn’s disease)
- Pyogenic granuloma-like lesion associated with topical retinoid therapy
- Pseudokaposiform reaction to Monsel’s solution
- Florid vascular proliferation as a needle biopsy effect in the breast
- Post-radiation vascular proliferations

### Table XVI. Histologically malignant tumors mistaken for benign.

- Inflammatory mesenchymal sarcomas (“pseudo-pseudotumors”) (e.g. inflammatory leiomyosarcoma, inflammatory fibrosarcoma, inflammatory malignant fibrous histiocytoma) versus myofibroblastic pseudotumors and tumefactive fibroinflammatory lesions
- **Low grade fibromyxoid sarcoma**
  - versus myxoid neurofibroma, perineurioma, desmoid fibromatosis, nodular fasciitis
- **Low grade myxofibrosarcoma**
  - versus myxoid neurofibroma, perineurioma, intramuscular / juxta-articular myxoma, superficial angiomyxoma
- **Low grade myofibroblastic sarcoma**
  - versus desmoid fibromatosis
- **Well differentiated lipoma-like liposarcoma**
  - versus ordinary lipoma, hibernoma
- **Well differentiated sclerosing type liposarcoma**
  - versus pleomorphic lipoma
- **Well differentiated inflammatory type liposarcoma**
  - versus inflammatory process
- **Well differentiated spindle cell (neural-like) liposarcoma**
  - versus benign peripheral nerve sheath tumor
- **Epithelioid sarcoma**
  - versus pseudogranulomatous inflammatory process (deep granuloma annulare, rheumatoid nodule, deep rheumatoid nodule, necrobiosis lipoidica, necrobiotic granuloma, fibrous benign tumors)
- **Subcutaneous panniculitis-like T cell lymphoma & granulomatous slack skin**
  - versus lupus profundus vs lymphocytic lobular panniculitis vs. atypical lymphocytic lobular panniculitis
- **Histiocytic sarcoma**
  - versus benign histiocytosis
- Non-mesenchymal tumors in special locations (low grade sarcomatoid carcinoma in the breast)
  - versus fibromatosis
Table XVII. Atypical variants of typical tumors.

A. By clinical features (the so-called “man from Istanbul” [127])

- Any typical superficial soft tissue tumor in deep location (e.g. deep juvenile xanthogranuloma – subcutaneous, intramuscular, intraneural and visceral forms [124, 125]; retroperitoneal spindle cell lipoma; retroperitoneal Pacinian neuroma; soft tissue deep and visceral [126, 127] benign fibrous histiocytoma; deep - subcutaneous dermatofibrosarcoma protuberans without dermal involvement; …)
- Any typical deep soft tissue tumor in very superficial location (e.g. cutaneous rhabdomyosarcoma; cutaneous – intradermal liposarcoma; Ewing’s sarcoma of the skin; adamantinoma of the pretibial soft tissue; …)
- Any typical tumor in heterotopic/unusual locations (e.g. fibrous hamartoma of infancy in hands or feet; non-midline / non-paraxial paraganglioma; subcutaneous/intravascular/visceral-organ based pyogenic granuloma; …)
- Kimura’s disease in non-Orientals
- “Extra-nodal” Castleman’s disease
- “Extra-nodal” Kimura’s disease
- “Extra-nodal” Rosai-Dorfman disease
- “Extra-nodal” dendritic cell and interdigitating cell sarcomas
- “Extra-abdominal” intra-abdominal desmoplastic round cell tumor (pleura, meninges, …)
- “Extra-orbital” (orbital) giant cell angiofibroma
- “Extra-nasopharyngeal” nasopharyngeal angiofibroma [128, 128a]
- “Extra-renal” renin-producing juxtaglomerular tumor
- “Non-acral” acral inflammatory myxohyaline tumor
- “Extra-acral” (acral) calcifying aponeurotic fibroma
- “Non-digital” (long bones) fibroosseous pseudotumors of the digits
- “Extra-digital” infantile digital fibromatosis
- “Extra-gastrointestinal” gastrointestinal stromal tumor
- “Extra-digit” infantile digital fibromatosis
- “Extra-mammary” mammary-type myofibroblastoma
- “Extra-neural” cerebellar-type hemangioblastoma
- “Extra-uterine” (parasitic) uterine leiomyoma
- “Extra-ulnar” (ulnar) cellular angiofibroma
- (Vulval) angiomyofibroblastoma in males (scrotum)
- (Vulval) angiomyofibroblastoma-like tumor in males (analogous to cellular angiofibroma)
- Aggressive angiomomyxoma (of the female pelvis) in males
- Nasopharyngeal angiofibroma (of the males) in females
- Juvenile xanthogranuloma in adults
- Infantile digital fibromatosis in adults
- Infantile myofibromatosis in adults
- Calcifying fibrous pseudotumor of childhood in adults
- Kaposiform (infantile) hemangioendothelioma in adults
- Ectopic meningioma
- Pediatric-type sarcomas in adults (including embryonal & botryoid rhabdomyosarcoma) PNET/EWS, angiomatoid malignant fibrous histiocytoma, pleomorphic fibrohistiocytic tumor [131]
- Spindle cell rhabdomyosarcoma in adults [132]
- Liposarcoma in children (questioned – debated)
- MPNST in children (questioned – debated)
- “Extra-medullary” myeloid tumors (lymph node, upper respiratory tract, female genital tract, soft tissue, body cavities …)
- “Extraosseous” plasmacytoma (upper respiratory tract, lymph node, soft tissue, skin, mediastinum, …)
- “Extraneuraxial” (retroperitoneal malignant) meningioma [133]
- “Extra-uterine” (retroperitoneal) endometrial stromal sarcoma [134]
- Heterotopic (retroperitoneal) germ cell tumor
- “Non-phosphaturic” phosphaturic mesenchymal tumor
B. By clinical behaviour
- Metastasizing benign fibrous histiocytoma
- Metastasizing benign uterine leiomyoma
- Metastasizing benign meningioma
- (Metastasizing benign mixed tumors of salivary glands)

C. By macroscopic features
- “Solid” aneurysmal cyst of soft tissue and bone
- “Non-protuberans” (atrophic, plaque-type) dermatofibrosarcoma protuberans
- Encapsulated intradermal dermatofibrosarcoma protuberans
- “Non-ossifying” ossifying fibromyxoid tumor

D. By cytological / histological features
- Cell size / cell type
  i. Desmoplastic small round cell tumor, large cell variant
  ii. Ewing's sarcoma, large cell (atypical) variant
  iii. Ewing's sarcoma, pleomorphic/anaplastic variant
  iv. Merkel cell tumor, large cell variant
  v. Anaplastic large cell lymphoma, small cell variant
  vi. Non-pleomorphic spindle-cell atypical fibroxanthoma
  vii. Juvenile xanthogranuloma without multinucleated giant cells
  viii. Plexiform fibrohistiocytic tumor without multinucleated giant cells
  ix. Tenosynovial giant cell tumor without giant cells

- Cell / matrix peculiarities
  i. Amelanotic melanoma
  ii. Non-lipidized juvenile xanthogranuloma (early phase)
  iii. Sclerotic lipoma
  iv. Fibrous spindle cell lipoma
  v. Extraskeletal myxoid chondrosarcoma without myxoid areas
  vi. Ewing's sarcoma, schlerosing variant

- Cell shape
  i. Spindle cell melanoma
  ii. Spindle cell lipoma
  iii. Spindle cell hemangiendothelioma
  iv. Leiomyomatous (spindle cell) rhabdomyosarcoma (both in children and in adults)
  v. Fibroma-like variant of epithelioid sarcoma
  vi. Ewing's sarcoma, spindle cell variant
  vii. Spindle cell lymphoma
  viii. Sarcomatoid plasmacytoma
  ix. Metastatic spindle cell (fibromatosis-like, sarcomatoid) carcinoma

- Architectural pattern
  i. Kaposi's sarcoma (nodular stage) without erythrocyte extravasation
  ii. Solid variant of alveolar rhabdomyosarcoma
  iii. Solid variant of aneuysmal bone cyst
  iv. Ewing's sarcoma with adamantanoma-like pattern

- Immunohistochemistry
  i. Sarcoma with partial epithelial differentiation (Ewing's sarcoma, fibroblastic dendritic reticulum cell sarcoma)
  ii. Any tumor with divergent/aberrant expression (e.g. keratins in melanoma, lymphoma, vascular neoplasms, smooth muscle neoplasms; neuroendocrine markers in sarcoma (chondrosarcoma with neuroendocrine differentiation);)
  iii. Any tumor with loss of specific antigen (e.g. S-100 protein and HMB-45 negative desmoplastic and non-desmo plastic melanoma, cytokeratin negative sarcomatoid carcinoma)
  iv. Non-germ cell tumors expressing germ-cell markers (e.g. PLAP expression in tumors with myogenic differentiation and desmoplastic small round cell tumor)
### Table XVIII. Cystic tumors and pseudotumors.

**A. Primarily cystic lesions**
- Cutaneous myxoid cyst
- Myxoid cyst of tendon sheath (ganglion)
- Nerve sheath ganglion
- Cavernous lymphangioma (cystic hygroma)
- Multicystic mesothelioma
- Lymphangioleiomyomatosis

**B. Lesions with secondary cystic or microcystic change**
- Fat necrosis
- Membranocystic fat necrosis / membranocystic panniculitis / membranous lipodystrophy (primary idiopathic, and secondary)
- Nodular-cystic-encapsulated fat necrosis (“mobile encapsulated lipoma”) 141
- Lipoma with fat necrosis (including membranous type)
- Cystic nodular fasciitis
- Myositis ossificans
- Ancient schwannoma
- Perineurioma
- Myxolipoma
- **Superficial angiomyxoma & juxtarticular myxoma**
- **Low grade fibromyxoid sarcoma & hyalinizing spindle cell tumor with giant rosettes**
- Ossifying fibromyxoid tumor (occasional)
- Giant cell tumor of soft tissue
- Angiomatoid fibrous histiocytoma
- Myxofibrosarcoma
- Myoid leiomyosarcoma
- Myoidliposarcoma
- (Extra-)gastrointestinal stromal tumor
- Aggressive angiomyxoma
- Ewing’s sarcoma of the skin (cystic spiradenoma-like tumor)
- Synovial sarcoma

**C. Pseudocystic pattern**
- Intravenous leiomyomatosis
- Necrotic sarcomas

### Table XIX. Acral tumors and pseudotumors.

- Rudimentary supernumerary digit
- Acquired digital fibrokeratoma
- Infantile digital fibromatosis
- **Superficial acral fibromyxoma**
- Acral myxoinflammatory fibroblastic sarcoma
- Cellular digital fibromas
- Giant cell tumor of tendon sheath – localized-type
- Glomus tumors
- **Sclerosing perineurioma**
- Hybrid schwannoma-perineurioma tumors 68
- Calcifying aponeurotic fibroma
- Chondroma
- **Fibroosseous pseudotumor of the digits**
- Lipofibromatous hamartoma of the nerve (with or without macrodactyly) 141a
- Nora’s lesion
- Fibroma of tendon sheath
- Fibromatosis (palmar: Dupuytren’s disease; plantar: Ledderhose’s disease; penile: Peyronie’s disease)
- Congenital & infantile fibrosarcoma
- **Clear cell sarcoma**
- **Epithelioid sarcoma** (conventional or “distal” type)
Table XX. Orbital tumors and pseudotumors.

- Neuromuscular choristoma / hamartoma
- Infantile myofibromatosis
- Periorbital granuloma annulare
- Nodular fasciitis
- Intravascular papillary endothelial hyperplasia (Masson's phenomenon)
- Amputation neuroma (following enucleation)
- Amyloid tumor (amyloidoma)
- Inflammatory pseudotumor
- Hemangioma
- Benign fibrous histiocytoma
- Spindle cell lipoma
- Pleomorphic lipoma
- Myolipoma
- Peripheral nerve sheath tumors
- Meningioma (*lipoblastic type* [142], ...)
- Granular cell tumor
- Giant cell angiofibroma
- Solitary fibrous tumor
- Hemangiopericytoma [143]
- Lipomatous hemangiopericytoma ([lipomatous solitary fibrous tumor] [144]
- Giant cell tumor (benign osteclastoma)
- Synovial sarcoma
- Liposarcoma
- Rhabdomyosarcoma (including sclerosing variant)
- Alveolar soft part sarcoma
- Mesenchymal chondrosarcoma
- Granulocytic sarcoma
- Leukemia / lymphoma
- Metastatic tumors (sarcoma, chordoma, carcinoma, myoepithelioma, carcinoid, ...)

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[142] References not provided in the text.
[143] References not provided in the text.
[144] References not provided in the text.
### Table XXI. Paratesticular soft tissue tumors and pseudotumors.

**A. Benign tumors and pseudotumors**

- (Adrenal rests)
- Extrarenal nephrogenic rests 145
- Angiokeratoma – Fordyce-type, solitary form
- Angiokeratoma – Fordyce type, multiple, iatrogenic form 146
- Smooth muscle hyperplasia of testicular adnexa
- Idiopathic scrotal calcinosis
- Mechanium periorchitis
- Sclerosing lipogranuloma
- Fibromatous periorchitis / nodular periorchitis
- Granulomatous lymphangitis of the scrotum and penis
- Granulomatous and nongranulomatous epididymitis
- Malakoplakia
- Mesothelial hyperplasia
- Perineal nodular induration in cyclists (3rd testicle / accessory testicle)
- Fibroma of testicular tunics
- Proliferative funiculitis
- Fibromatosis (of spermatic cord)
- Traumatic neuroma (of the penis)
- Lipoma
- Liposarcoma
- Leiomyoma – ordinary (mainly scrotal) & scrotal bizarre leiomyoma
- Myoepitheloma (penis)
- Rhabdomyoma
- Hemangioma
- Neurofibroma
- Fibrous hamartoma of infancy 147
- Granular cell tumor
- Adenomatoid tumor
- Inflammatory myofibroblastic tumor
- Calcifying fibrous pseudotumor
- Angiomyxolipoma
- Angiomyofibroblastoma
- Angiomyofibroblastoma-like tumor in males (analogous to vulval cellular angiofibroma)
- Aggressive angiomyxoma
- Spleno-gonadal fusion
- Solitary fibrous tumor (of the spermatic cord) 148

**B. Malignant tumors**

- Melanotic neuroectodermal tumor of infancy
- Leiomyosarcoma (paratesticular, and penile)
- Fibrosarcoma
- Rhabdomyosarcoma (including the spindle cell variant of children)
- Liposarcoma (mainly well differentiated)
- Liposarcoma with smooth muscle differentiation
- Malignant fibrous histiocytoma
- Epithelioid sarcoma (penis)
- Synovial sarcoma (perineum, penis)
- Osteosarcoma (penis)
- Well differentiated papillary mesothelioma of the tunica vaginalis / malignant mesothelioma of the tunica vaginalis 149
- Paratesticular desmoplastic small round cell tumor
Table XXII. Vulvo-vaginal soft tissue tumors and pseudotumors 150.

- Fibromatosis
- Nodular fascitis
- Fibroepithelial stromal polyp (bland type; pseudosarcomatous) 50
- Childhood asymmetric labium majus enlargement (aka, prepubertal vulval fibroma) 151 152
- Spindle cell lipoma
- Pleomorphic lipoma
- Cellular angiofibroma
- Dermatofibrosarcoma protubersans
- Acquired vulvar lymphangioma (secondary to Crohn’s disease)
- Vulval angiokeratoma (Fordyce type) 155
- Angiomyofibroblastoma
- Angiomyofibrosarcoma (malignant angiomyofibroblastoma)
- Superficial angiomyxoma
- Aggressive angiomyxoma
- Post-hysterectomy Fallopian tube prolapse 154 155
- Myoepithelioma – “true” myoepithelioma
- Adenomyoepithelioma
- “True” mixed tumor of the vulva
- So-called “mixed tumor” of vagina (spindle cell epithelioma) 156 157
- Lipoblastoma-like tumor
- Spindle cell / pleomorphic lipoma
- Atypical lipomatous tumor 158
- Smooth muscle tumors (any type, including the clear-cell epithelioid leiomyoma of the round ligament 158a)
- Genital rhabdomyoma
- Paraganglioma 159
- Perivascular epithelioid cell neoplasms 160
- Epithelioid sarcoma
- Synovial sarcoma
- Extrarenal malignant rhabdoid tumor
- Melanoma & malignant blue nevus
- Melanoma with osteocartilaginous differentiation 161
# Table XXIII. Aerodigestive tract lesions (involving the orifices).

## A. Nasal cavity
- Mycobacterial spindle cell pseudotumor
- Amyloid tumor (amyloidoma)
- Nasal polyps with atypical stromal cells – a pseudosarcomatous lesion
- Congenital nasal hemangiopericytoma / infantile myofibromatosis
- Nasal chondromesenchymal hamartoma in children
- Nasal chondromesenchymal hamartoma in children and adults
- Meningioma
- Nasal glioma – so-called (nasal cerebral heterotopia or sequestered encephalocele)
- Paraganglioma
- Olfactory neuroblastoma
- Sinonasal teratocarcinosarcoma ("mixed olfactory neuroblastoma-craniofibroma") / teratocarcinosarcoma (olfactory neuroblastoma with glandular differentiation)
- Extramedullary plasmacytoma
- Solitary fibrous tumor
- Hemangiopericytoma-like tumor ("sinonasal-type hemangiopericytoma")
- Nasopharyngeal and extranasopharyngeal angiofibroma
- Smooth muscle tumors
- Angiomyolipoma
- Spindle cell myoepithelioma
- Undifferentiated small cell tumor
- Alveolar rhabdomyosarcoma (paranasal sinuses)
- Synovial sarcoma
- Malignant melanoma

## B. Oral cavity
- Rhabdomyomatous mesenchymal hamartoma
- Fibrous hamartoma of infancy
- Congenital granular cell epulis
- Amyloid tumor (amyloidoma)
- Neuroma
- Schwannoma
- Granular cell tumor
- Rhabdomyoma
- Multifocal rhabdomyoma
- Ectomesenchymal chondromyxoid tumor of the tongue
- Myoepithelioma
- Fibrolipoma
- Ordinary lipoma
- Spindle cell lipoma
- Pleomorphic hyalinizing angiectatic tumor
- Peripheral myxoma
- Solitary fibrous tumor
- Congenital hemangiopericytoma / infantile myofibromatosis
- Epithelioid hemangioperichyma
- Leiomyosarcoma
- Liposarcoma
- Embryonal rhabdomyosarcoma
- Malignant fibrous histiocytoma
- Clear cell sarcoma
- Lingual alveolar soft part sarcoma
- Low-grade malignant Triton tumor
- Synovial sarcoma
- Ordinary melanoma
- Melanoma with osteocartilaginous differentiation

## C. Anal canal
- CD34+ fibroepithelial polyp
- Aggressive angiomyxoma
- Schwannoma & neurofibroma
- Leiomyoma
- Leiomyosarcoma
- Gastrointestinal stromal tumor
Table XXIV. Intranodal primary tumors & pseudotumors (other than hematolymphoid).

- Nodular fasciitis (of the capsule)
- Amyloid tumor
- Deciduosis
- Lipomatosis
- Angiomyolipoma
- Lymphangioleiomyomatosis
- Inflammatory pseudotumor
- Mycobacterial spindle cell pseudotumor
- Bacillary angiomatosis
- Rosai-Dorfman disease
- Castleman’s disease (hyaline-vascular type)-related tumors (follicular dendritic cell tumor, Kaposi’s sarcoma) and pseudotumors (angiomyolipomatous hamartoma)
- Angiomyomatous hamartoma
- Lymphangioma
- Angiolymphatic angiodyplasia
- Vascular transformation of sinuses
- Nodular spindle-cell vascular transformation of lymph nodes
- Nodal angiomia (common type – capillary, cavernous, lobular, cellular)
- Epitheloid hemangioma
- Epitheloid hemangioendothelioma
- Spindle and epitheloid hemangioendothelioma
- Polymorphous hemangioendothelioma
- Angiosarcoma
- Kaposi's sarcoma
- Leiomyoma
- Palisaded myofibroblastoma
- Plexiform neurofibroma
- Myelolipoma
Table XXV. Mammary stroma spindle cell tumors and tumor-like lesions.

A. Monomorphic spindle cell tumors and tumor-like lesions of specialized mammary stroma

- Pseudoangiomatous stromal hyperplasia
- Benign spindle cell tumor, NOS
- Spindle cell lipoma-like tumor
- Solitary fibrous tumor
- Myofibroblastoma
- Combined spindle cell tumor (two of the above: e.g., myofibroblastoma plus solitary fibrous tumor areas or spindle cell lipoma-like areas)
- Periductal stromal tumor
- Stromal sarcoma

B. Monomorphic spindle cell tumors and tumor-like lesions of non-specialized mammary stroma

- Nodular fasciitis
- Lipoma
- Muscular hamartoma
- Benign fibrous histiocytoma
- Fibromatosis
- Leiomyoma
- Inflammatory myofibroblastic tumor (benign, low-grade)
- Spindle cell adult type juvenile xanthogranuloma
- Angiomyolipoma
- Fibrosarcoma
- Myofibrosarcoma
- Leiomyosarcoma
- Peripheral nerve sheath tumor (benign and malignant)
- Monophasic fibrous synovial sarcoma
- Hemangiopericytoma
- Mammary NOS-type sarcoma with CD10 expression and features of myoepithelial differentiation

C. Spindle cell myoepithelioma

- Myoepithelioma

D. Spindle cell epithelial tumors

- Metaplastic carcinoma – biphasic and monophasic
- Monophasic low-grade spindle cell fibromatosis-like carcinoma
- Monophasic high-grade spindle cell (sarcomatoid) carcinoma (including a subset showing myoepithelial differentiation)
### Table XXVI. Infantile-juvenile tumors and pseudotumors.

**A. Hamartomas – Developmental lesions**
- Connective tissue nevi
- Palmar cutaneous hamartoma
- Nevus lipomatosus superficialis
- Cutaneous neural heterotopia
- Cutaneous ganglion cell choristoma
- Hamartomatous neuroma combined with heterotopic ganglion cells
- Congenital neural hamartoma (“fascicular schwannoma”)
- Multiple cutaneous neuromuscular choristomas
- Pleomorphic neuromuscular hamartoma of the subcutis (superficial equivalent of deep neuromuscular hamartoma or benign Triton tumor)
- Congenital neurovascular hamartoma of the skin
- Fibrous hamartoma of infancy
- Congenital smooth muscle hamartoma of the skin
- Genital smooth muscle hamartoma
- Rhabdomyomatous mesenchymal hamartoma
- Striated muscle hamartoma
- Neuromuscular hamartoma / choristoma (benign Triton tumor)
- **Neural fibrolipomatous hamartoma**
- Ectopic meningothelial hamartoma of the scalp
- Sequestrated (rudimentary) meningocele
- Sacro-coccygeal (“myxopapillary”) ependymal rests
- Meningo-gial nodule of the buttock
- Soft tissue gliomatosis in a non-midline location
- Neurocristic cutaneous hamartoma
- Nasal chondromesenchymal hamartoma
- Mesenchymal hamartoma of the chest wall
- Lipomatous and angiomatic lesion associated with occult spinal dysraphism

**B. Benign tumors and pseudotumors**
- Granular (gingival) cell tumor
- Hemangioma
- Lymphangioma
- Infantile fibromatosis, diffuse and desmoid type
- Infantile digital fibromatosis
- Infantile myofibromatosis
- Infantile hemangiopericytoma
- Juvenile hyaline fibromatosis / infantile systemic hyalinosis / and Winchester syndrome
- Proliferative fasciitis
- Proliferative myositis
- Cranial fasciitis
- Fibrous umbilical polyp (fasciitis-like proliferation of infancy)
- Umbilical pseudotumors (keloid/hypertrophic scar, umbilical polyp/granuloma with urachal, omphalomesenteric, or other remnants)
- Lipoblastoma / lipoblastomatosis
- Lipofibromatosis
- Myositis ossificans progressiva (fibrodyplasia)
- Benign and atypical fibrous histiocytoma
- Giant cell fibroblastoma
- Juvenile xanthogranuloma
- Fetal rhabdomyoma
- Fetal rhabdomyoma, cellular-type
- Juvenile rhabdomyoma
- Cellular capillary hemangioma
- Intramuscular hemangioma
- Kaposi-like infantile hemangioendothelioma
- Spindle cell hemangioendothelioma
- Congenital or infantile hemangioendothelioma
- Congenital giant cell angioblastoma
- Nasopharyngeal and extra-nasopharyngeal angiofibroma
- Congenital and childhood plexiform (multinodular) cellular schwannoma
### C. Intermediate and malignant tumors

- Congenital myofibromatosis (visceral involvement)
- Congenital (infantile) fibrosarcoma
- Kaposi’s sarcoma (HIV-negative, peripartum HHV-8 infection)
- Lymphangioendotheliomatosis
- Primitive myxoid mesenchymal tumor \(^{202a}\)
- Pigmented (melanotic) neuroectodermal tumor of infancy (maxilla, mandible, mediastinum, skull, epididymis, soft tissue of extremities)
- **Inflammatory pseudotumor – Inflammatory myofibroblastic tumor**
- Plexiform fibrohistiocytic tumor
  - Pediatric dermatofibrosarcoma protuberans (rare; occasionally congenital)
  - Pediatric pigmented dermatofibrosarcoma protuberans \(^{200b}\)
  - Pediatric leiomyosarcoma
- Endovascular papillary angioendothelioma (Dabska’s tumor)
- Ewing’s sarcoma
  - Congenital Ewing's sarcoma
  - Primitive neuroectodermal tumor
  - Rhabdomyosarcoma (embryonal, alveolar; rarely congenital)
  - **Infantile rhabdomyosarcoma**
  - Synovial sarcoma (even congenital)
  - Congenital angiomatoid malignant fibrous histiocytoma
  - Congenital synovial sarcoma
  - Congenital extraskeletal embryonal chondrosarcoma
  - Alveolar soft part sarcoma
  - Extrarenal malignant rhabdoid tumor
  - Congenital disseminated extrarenal malignant rhabdoid tumor
  - Angiomatoid (malignant) fibrous histiocytoma
  - Pediatric leiomyosarcoma of soft tissue (rare)
  - Pediatric epithelioid sarcoma (rare) \(^{201}\)
  - Pediatric malignant peripheral nerve sheath tumor (rare) \(^{202}\)
  - Pediatric malignant vascular tumors \(^{205}\)
  - Liposarcoma (exceptional)
  - Pediatric clear cell sarcoma (of tendons and aponeurosis) \(^{204}\)
  - Primitive undifferentiated sarcoma
  - Congenital leukemia cutis
  - Cutaneous neonatal neuroblastoma
  - Soft tissue sarcoma as a second malignant neoplasm \(^{205}\)
Table XXVII. Lesions as sentinel of clinical entities (see also Table XXVIII).

- Keloid (acne vulgaris; ear-piercing; previous surgical intervention or thermal injuries)
- Membranocystic fat necrosis / membranocystic panniculitis / membranous lipodystrophy (secondary form - mainly lower leg venous insufficiency in obese women, erythema nodosum, morphea profunda, lupus panniculitis, dermatomyositis, subcutaneous T-cell lymphoma, factitial ulcer, … others)
- Nodular panniculitis (Weber-Christian disease; alpha-1-antitrypsin-deficiency; Rothmann-Makai panniculitis; pancreatic disease)
- Massive localized lymphedema (morbid obesity)
- Polvlyvinylpyrrolidone granuloma (polymer used as plasma expander in a previous surgical intervention or as vehicle for intravenous medications)
- Pseudosclerodermatous panniculitis of the breast (previous radiation therapy for breast cancer)
- Pseudolipoma
- Folded skin with lipomatous nevus ("Michelin-tire baby")
- Multiple nevus lipomatous cutaneous superficialis (Hoffman-Zurhelle disease)
- Granular histiocytic granuloma (local previous site of surgery)
- Scleredema (maturity-onset diabetes mellitus)
- Xanthogranulomatous pseudotumor (Erdheim-Chester disease)
- Superficial fibromatoses (association of palmar with plantar and/or penile forms)
- Multiple desmoids tumors (familial infiltrative fibromatosis)
- Myofibroma (myofibromatosis – bone, and visceral organ involvement; possible familial occurrence)
- Kimura’s disease (minimal change/membranous glomerulopathy nephrotic syndrome, peripheral blood eosinophilia; elevated serum IgE)
- Paraffinoma – oleogranuloma (cosmetic purposes | breast of females, breast of transsexual males, eyelids, lips, male genitalia – scrotum, penis; pseudotherapeutic and therapeutic procedures (scalp for baldness, sinonasal surgery and medication, intraleral injection of oil based chemotherapeutic agents)
- Gingival pyogenic granuloma (granuloma gravidarum) (pregnancy)
- Soft tissue and cutaneous deciduosis (pregnancy; occasionally in non-pregnant women)
- Leiomyomatosis peritonealis disseminata (excess exogenous or endogenous sex steroids, pregnancy, endometriosis)
- Extramedullary hematopoiesis (anemia of various origin, chronic myeloproliferative disorders)
- Extramedullary myeloid tumor (acute or chronic myelogenous leukemia)
- Sclerosing extramedullary hematopoietic tumor (chronic myeloproliferative disorders)
- Extramedullary plasmacytoma (full-blown multiple myeloma)
- Cytophagic histiocytic panniculitis (subcutaneous T-cell lymphoma)
- Angiosarcoma (preexisting lymphedema, previous local irradiation, local lymphedema, …)
- Atypical vascular lesion of skin of the breast (previous irradiation)
- Mucosal, lymph nodal and/or visceral kaposi’s sarcoma (immunosuppression, AIDS)
- Verruga peruana / bacillary angiomatosis (Carrion disease / AIDS)
- Acquired progressive lymphangioma (irradiation, trauma, …)
- Epithelioid hemangioma (multicentricity)
- Multicentric reticulohistiocytosis (paraneoplastic disease)
- Xanthoma – various types (e.g. tendinous xanthoma: disorders of lipid metabolism)
- Plexiform xanthomatous tumor (hyperlipidemia)
- Calcinosis circumspecta and calcinosis universalis (scleroderma, dermatomyositis, Raynaud disease, hyperparathyroidism, …)
- Ectopic calcifications (hypercalcemia-associated conditions: milk-alkal-syndrome or Burnett syndrome, hypervitaminosis D, tumoral bone metastases) and calciphylaxis (associated with secondary hyperparathyroidism / chronic renal failure)
- (Idiopathic) tumoral calcinosis (unassociated with hypercalcemia: sometimes hyperphosphatemia-associated; sometimes familial)
- (Secondary) tumoral calcification-like lesion (associated with chronic renal failure and secondary hyperparathyroidism)
- Soft tissue calcifications and osteomas (Albright hereditary osteodystrophy)
- Cutaneous and subcutaneous myxoma (Carney’s complex)
- Pretibial myxedema (hyperthyroidism)
- Dermatofibrosarcoma protuberans (familial form)
- Multiple lipomas (familial form)
- Multiple spindle cell lipomas (familial form)
- Multiple angiolipomas (familial form)
- Symmetric lipomatosis of the head and neck (Madelung disease or Launois-Bensaude syndrome)
- Asymmetrical lipomatosis with “buffalo hump” (steroid lipomatosis - Cushing’s disease)
- Asymmetrical lipomatosis with wasting face and limbs (iatrogenic HIV lipodystrophy)
- Pelvic lipomatosis (mostly affecting blacks)
- Adiposis dolorosa (Dercum’s disease)
- Multiple schwannoma (schwannomatosis)
- Multiple cutaneous chondromas (familial form)
- Amyloid tumor (plasma cell dyscrasia; chronic inflammatory/autoimmune diseases; chronic hemodyalisis; others)
- Pseudoneoplastic parathyromatosis following iatrogenic implantation (hyperparathyroidism)
- Phosphaturic mesenchymal tumor (oncogenic osteomalacia/rickets)
- Idiopathic retroperitoneal fibrosis – Ormond’s disease (association with other fibrosclerotic disorders, e.g. Riedel thyroiditis, primary sclerosing cholangitis, sclerosing mediastinitis, inflammatory pseudotumor of the orbit, autoimmune pancreatitis)
- Tamm-Horsfall protein deposition / “retroperitoneal urinary precipitates” (past urine extravasation)
- Hemangiopericytoma & solitary fibrous tumor (hypoglycemia)
- (Retroperitoneal) extrarenal juxtaglomerular cell tumor (secondary hyperaldosteronism – hypertension, hypokalemia)
- Angiomyolipoma & lymphangioleiomyomatosis (tuberous sclerosis complex)
- Malignant Triton tumor and epithelioid malignant peripheral nerve sheath tumor (neurofibromatosis I)
- Paragangliomas (both head and neck and retroperitoneal) – either alone (PGL1, PGL2, PGL3, PGL4 syndromes) or as a component of a multiple tumor syndrome (von Hippel-Lindau disease, neurofibromatosis type 1, MEN-2b, Carney’s triad, Carney’ and Stratakis’ syndrome or in association with gastrointestinal stromal tumor)
- Gastrointestinal stromal tumor (familial form, and syndromic form in association with lentigines and mast cell tumors)
- Multifocal (EBV-associated) smooth muscle tumors (post-transplantation, congenital immunodeficiencies)
- Multifocal visceral smooth muscle tumors (immunocompromised HIV-patients)
- Myxoid embolus (cardiac myxoma)
- Sarcomatous embolus (great vessel sarcoma)
Table XXVIII. Lesions as sentinel of clinical syndromes (see also Table XXVII).

- Angiofibroma of the face (tuberous sclerosis complex)
- Periungual, subungual, and gingival angiofibroma (tuberous sclerosis complex; Gardner syndrome)
- Fibrofolliculoma (Birt-Hogg-Dube syndrome) 217
- Fibrous papule of the face (multiple hamartoma syndrome; Cowden's syndrome)
- Cutaneous myxoma (mainly of external ear location) (LAMB / NAME / Carney’s complex)
- Gardner-associated fibroma (Gardner syndrome; familial infiltrative fibromatosis; desmoid fibromatosis)
- Nuchal-type fibroma (diabetes mellitus; Gardner syndrome)
- Desmoid tumor (familial adenomatous polyposis / Gardner syndrome)
- Nasopharyngeal angiofibroma (familial adenomatous polyposis / Gardner syndrome)
- Keloids (Rubinstein-Taybi syndrome; Ehlers-Danlos syndrome)
- Gingival fibromatosis (Jimmiren-Laband syndrome; Ramon syndrome; Klippel-Trenaunay syndrome; Cowden’s syndrome; prune-belly syndrome)
- Juvenile hyaline fibromatosis and infantile systemic hyalinosis (Winchester syndrome)
- Gingival fibromatosis with juvenile hyaline fibromatoses (Murray-Puretic-Drescher syndrome)
- Penile fibromatosis (Cogan syndrome)
- Cerebriform superficial soft tissue fibrous proliferation (Proteus syndrome)
- Multiple lipomas (Proteus syndrome)
- Multiple intramuscular myxoma plus polyostotic fibrous dysplasia (Mazabraud’s syndrome)
- Lymphangioma (Turner syndrome)
- Lipomas and hemangiomas (Bannayan-Ruvalcaba-Riley syndrome; Cowden syndrome)
- Multiple lipoma (Bannayan-Zonana syndrome; Frolich syndrome; Proteus syndrome)
- Kaposiform hemangioendothelioma (Kasabach-Merritt syndrome)
- (Large) cavernous hemangioma (Kasabach-Merritt syndrome)
- Cutaneous and gastrointestinal venous malformations (blue rubber bleb nevus syndrome)
- Embryonal rhabdomyosarcomas (Beckwith-Wiedemann syndrome / Exomphalos-macroglossia-gigantism syndrome)
- Soft tissue sarcomas (retinoblastoma syndrome)
- Soft tissue sarcomas (Werner syndrome)
- Myogenic sarcomas (Rubinstein-Taybi syndrome)
- Pedal hemangioma (Turner syndrome)
- Hemangiomatosis (Klippel-Trenaunay syndrome)
- Angiomatous lesions (Sturge-Weber syndrome; Klippel-Trenaunay syndrome; Parkes-Weber syndrome; Rendu-Osler-Weber disease)
- Multiple hemangiomas and chondromas (Maffucci’s syndrome)
- Spindle cell hemangioma (Maffucci’s syndrome – occasional)
- Angiosarcoma, chondrosarcoma (Maffucci’s syndrome)
- Multiple glomus tumors and/or multiple glomus body hamartomas (congenital, familial)
- Glomeruloid hemangioma (POEMS / Crowe-Fukase / PEP / Takatsuki syndrome)
- Hemangioblastoma of soft tissue (von Hippel-Lindau syndrome)
- Lymphangioleiomyomatosis / soft tissue PEC-oma (tuberous sclerosis complex)
- Major and/or minor (2 or more) features of tuberous sclerosis complex plus autosomal dominant polycystic kidney disease (adjacent gene syndrome TSC2/ADPKD1 217a)
- Lymphangiosarcoma (Stewart-Treves syndrome) 218
- Mucosal neuromas (hypertrophied corneal nerves, bumpy lips, nodular tongue in MEN-2b)
- Plexiform neurofibroma and multiple neurofibromas (neurofibromatosis type I) 216a
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- Fetal rhabdomyoma (nevoid basal-cell carcinoma syndrome – Gorlin-Goltz syndrome) 219 220
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### Table XXIX. Embryological rests or normal anatomic structures potentially mistaken for tumors.

#### A. Normal embryological rests in soft tissue
- Branchial arch-derivatives (may mimic soft tissue chondroma)
- Branchial pouch-derivatives (may mimic metastatic carcinoma, ...)
- Supernumerary breast
- Lumbosacral ectopic nephrogenic rest (may mimic metastatic or ectopic Wilms' tumor, monodermal teratoma, ...) associated and unassociated with spinal dysraphism
- Sacrococcygeal ependymal rests (resembles ependymal tumors, metastatic papillary carcinoma, ...)
- Odontogenic epithelial rests

#### B. Normal anatomic structures in appropriate locations
- Dental papilla (resembling myxoma)
- Accessory tragus-elastic cartilage (resembling chondroma)
- Glomus coccygeum (resembling glomus tumor)
- Organ of Zuckerkandl hypertrophy (resembling paraganglioma)
- Carotid body hyperplasia (resembling paraganglioma)
- Chievitz organ (resembling metastatic carcinomas)
- Pacinian corpuscle hyperplasia (resembling Pacinian neuroma / Pacinian neurofibroma / Pacinian perineuroma / onion-bulb neuropathies)

#### C. Normal anatomic structures in inappropriate locations
- Dermal thymus (brachio-ocular-facial syndrome) (may mimic thymoma)
- Gial heterotopia (may mimic glioma)
- Hamartoma of the scalp with ectopic meningothelial elements / rudimentary meningocele (may mimic various neoplasms, e.g. angiosarcoma, meningioma, ...)
- Extramedullary hemopoiesis in normal organs (soft tissue, breast, lymph node, brain, ...) (may mimic lympho- or myelo-proliferative processes)
- Extramedullary hemopoiesis in vascular tumors (hemangioblastoma, spindle cell lipoma, pyogenic granuloma, ...)
- Subcutaneous splenosis (resembles hematologic proliferations)
- Langerhans' cell islets in adipose peripancreatic tissue (resembles metastatic carcinoid)
- Discontinuous splenogonadal fusion
- Any other adult tissue ectopically located in soft tissue (e.g. post-traumatic splenosis in the abdomen, autografted parathyroid in the forearm...
**Table XXX. Categorization** I-IV of soft tissue tumors and its relevance to management.

1 Pathologic categorization is defined on the basis of the clinical behaviour of each entity.
2 This clinically-based categorization of soft tissue tumors broadly corresponds to that found in specialized standard textbooks\(^{235-237}\) and major or specific publications.
3 The range of metastatic rates given within each category/group is an approximation gauged from the literature and from the authors’ experience, and – although it reflects the basis for the categorization – should be interpreted as approximate guide only.
4 Sarcomas are stratified according to a 3-tiered prognostic scheme, intended as a “working formulation” and not as a grading system along the lines of existing traditional grading schemes such as FNCLCC (see below). This stratification conveys notable differences in metastatic rate.

### Category I. Clinically benign tumors

<table>
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<tr>
<th>Metastatic rate of nearly 0%. Local excision is usually curative</th>
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| Dermatofibroma (**including variants** – clear cell, epithelioid, sclerosing, myxoid, keloidal, palisading, lipidized, ...) & cutaneous benign fibrous histiocytoma\(^{\wedge}\), fibroma (**including variants** – sclerotic, pleomorphic, *nuchal-type*, Gardner-associated type, ...), fibroma of tendon sheath, giant cell tumor of tendon sheath (localized-type), superficial fibromatosis (*prone to recurrence*)\(^{\wedge}\), superficial fibromatosi**desmoid tumor (*prone to recurrence*)\(^{\wedge}\), nasopharyngeal angiofibroma (*prone to recurrence*), desmoplastic fibroblastoma (collagenous fibroma), mammary-type fibroblastoma, giant cell fibroblastoma (*may recur*), intranodal palisaded myofibroblastoma, dermatomyofibroma, myofibromatosis infantile-type (*“infantile hemangio- pericytoma*), intramuscular myxoma (*including the cellular variant*, calcifying fibrous pseudotumor, calcifying aperoneurotic fibroma (*prone to recurrence*), juxtaarticular myxoma, cutaneous myxoma, superficial angiomylipoma with and without epithelial elements (*prone to recurrence*)\(^{238}\), “aggressive” angiomyxoma of the pelvis (*prone to recurrence*), vulvovaginal and inguinoperineal angiomyxofibroblastoma, nasopharyngeal angiofibroma (*prone to recurrence; locally aggressive*), vulval cellular anaplasia, basaloid squamous cell carcinoma (*including the cellular variant*, calcifying fibrous pseudotumor, calcifying aperoneurotic fibroma (*prone to recurrence*), vulvar cellular anaplasia, nasal squamous cell carcinoma (*including the cellular variant*, calcifying fibrous pseudotumor, calcifying aperoneurotic fibroma (*prone to recurrence*), vulvar cellular anaplasia, basaloid squamous cell carcinoma (*including the cellular variant*, calcifying fibrous pseudotumor, calcifying aperoneurotic fibroma (*prone to recurrence*), vulvar cellular anaplasia, nasal squamous cell carcinoma (excluding the psammomatous melanotic type), neurofibroma (*may recur*), schwannoma/perineurioma, usual granular cell tumor (*may recur*), neurothekeoma (myxoid-type, cellular-type), soft tissue chondroma, fibrochondroma, myxochondroma, osteoma, and osteochondroma (*including variants* – multiple, familial), extraarticular synovial chondromatosis, soft tissue oncycytoma (*little experience*), hyaline cell-rich chondroid syringoma, ectopic hamartomatous thymoma\(^{\sqrt{\wedge}}\), myelolipoma.

\(^{\wedge}\) Some variants – atypical, cellular, aneurysmal, deepseated – may recur and anecdotal metastatic cases\(^{135}\) are also on record. \(^{\wedge}\) May undergo malignant change. \(^{\wedge}\) 2 metastasizing cases on record\(^{239,240}\). \(^{\wedge}\) 1 case of malignant transformation\(^{241}\). \(\sqrt{\wedge}\) Malignant myopericytomas likely correspond to malignant peripheral nerve sheath tumors with perineural differentiation\(^{239,241}\). \(\sqrt{\wedge}\) 1 case of malignant transformation\(^{252}\).

Note: tumors “*prone to recurrence*” might be also placed in Group II (see footnote there).
**Category II. Clinically intermediate** (borderline) tumors

- Metastasis can rarely occur: metastatic rate of < 5%.
- Prone to recurrence if incompletely excised: attention must be paid to margins.
- Tumor may transform or dedifferentiate to group III (below).

Atypical fibroxanthoma (laryngeal variant — spindle cell non-pieomorphic, pigmented or aneurysmal, clear cell, granular cell, osteoclastic giant cell, ...), "some" cutaneous and subcutaneous "benign" fibrous histiocytomas (histology-dependent — atypical, cellular, aneurysmal, site-dependent — deep-seated, head & neck area; previous excision — recurring lesions)¹³,²⁵,²⁶, dermatofibrosarcoma protuberans² (classic-type, Bednar/pigmented type — any site, any age, any variant), hybrid dermatofibrosarcoma protuberans/giant cell fibroblastoma,plexiform fibrohistiocytic tumor, giant cell angiofibroma, solitary fibrous tumor (including lipomatous hemangiopericytoma), atypical lipomatous tumor, angiomatoid fibrous histiocytoma (aka, angiomatoid malignant fibrous histiocytoma), ossifying fibromyxoid tumor, inflammatory myofibroblastic tumor ("inflammatory pseudotumor"), inflammatory fibrosarcoma, pleomorphic hyalinizing angiectatic tumor — hemosiderotic fibrohistiocytic lipomatous tumor (prone to recurrence)²⁵⁴, myxoinflammatory fibroblastic sarcoma, deep-seated or large sized benign appearing smooth muscle tumors of soft tissue²⁴²,²⁴³, well-differentiated liposarcoma (lipoma-like variant; sclerosing variants), "atypical" glomus tumors (deep-seated only / superficially located & mitotically active only / large-sized only)²⁵⁵, endothelial neoplasms (Dabska's tumor or papillary intralymphatic angioendothelioma, retiform hemangiendothelioma, composite hemangiendothelioma, polymorphous hemangiendothelioma, giant cell angiofibroma, Kaposis's sarcoma²⁴⁵,²⁴⁶, giant cell tumor³ (skin, soft tissue, breast)²⁵⁶, phosphaturic mesenchymal tumor⁴ (excluding atypical variants), superficial low-grade fibromyxoid sarcoma (may recur)²⁵⁷, diffuse-type tenosynovial giant cell tumor ("pigmented villonodular synovitis/bursitis") (prone to recurrence)⁵, soft tissue mixed tumor, myoepithelioma, parachordoma.

³⁵ "Intermediate" is a term which is also used for tumors which can recur but do not metastasize; these are also (more appropriately) termed locally aggressive. ⁶ May undergo sarcomatous change.

**Category III (X, Y, Z groups). Clinically malignant tumors**

### Group III-X. Low-grade malignancy — metastatic rate of 5-10%.

Infantile/congenital fibrosarcoma, well-differentiated (fibromatosis-like / Broders gr. I) fibrosarcoma, low-grade fibromyxoid sarcoma¹ (including its variant hyalinising spindle cell tumor with giant rosettes), gr. I malignant fibrous histiocytoma ("low-grade" myxofibrosarcoma), angiomatoid (malignant) fibrous histiocytoma, cutaneous leiomyosarcoma, usual / well differentiated solitary fibrous tumor, low-grade myofibrosarcoma, well-differentiated leiomyosarcoma, leiomyosarcoma in children, well-differentiated liposarcoma with leiomyosarcomatous differentiation (w.d. lipolementosarcoma), spindle cell liposarcoma, "low-grade" dedifferentiated liposarcoma⁷, psamnomatous melanotic schwannoma, low-grade malignant peripheral nerve sheath tumor, well-differentiated (gr. I) chondrosarcoma, low-grade angiosarcoma²⁴⁵,²⁴⁶.

⁸ Ungradable according to some.

⁹ Note: here are two tiered grading systems: one is 2-tiered (low- and high-grade) and the other is 3-tiered (low-, intermediate-, and high-grade).

### Group III-Y. Intermediate-grade malignancy — metastatic rate 10-30%

Fibrosarcoma arising in dermatofibrosarcoma protuberans (classic-type, Bednar-type), moderately differentiated conventional fibrosarcoma, solitary fibrous tumor (atypical, malignant), "true" well-differentiated malignant hemangiopericytoma²⁵¹, gr. II myxoid malignant fibrous histiocytoma ("high-grade" myxofibrosarcoma), storiform malignant fibrous histiocytoma, conventional myxoid liposarcoma, conventional leiomyosarcoma, myxoid leiomyosarcoma, granular cell leiomyosarcoma (little experience), spindle cell embryonal rhabdomyosarcoma ("leiomyomatous variant"), moderately differentiated malignant peripheral nerve sheath tumor, conventionally dedifferentiated liposarcoma, malignant mesenchymoma, epithelioid hemangioendothelioma, "low-grade" extraskeletal myxoid chondrosarcoma²⁶² and its analogous variant with neuroendocrine differentiation, soft tissue follicular dendritic cell sarcoma.

⁵ we have used a 2-tiered grading system for myxoid malignant fibrous histiocytoma (Angerwall’s myxofibrosarcoma). ⁷ Classic histologies correlate with the lower end of metastatic rate of the category, while atypical features correlate with the upper end of the metastatic prognostic spectrum.
Group III-2. High-grade malignancy – metastatic rate > 30%.

Poorly differentiated conventional fibrosarcoma, sclerosing epithelioid fibrosarcoma, high grade myxoid/round cell liposarcoma, epithelioid liposarcoma, pleomorphic liposarcoma, some mixed-type liposarcoma (myxoid/round cell and pleomorphic liposarcoma), pleomorphic myofibrosarcoma, “true” conventional malignant hemangiopericytoma, poorly differentiated leiomyosarcoma, pleomorphic leiomyosarcoma, leiomyosarcoma, pleomorphic leiomyosarcoma, leiomyosarcoma with prominent osteoclastic giant cells, epithelioid leiomyosarcoma, inflammatory leiomyosarcoma, rhabdomyosarcoma (any type – embryonal, alveolar, pleomorphic, angiosarcoma) (any type – conventional, epithelioid, poorly differentiated), intimal sarcoma, glomangiosarcoma, malignant peripheral nerve sheath tumor (poorly differentiated, epithelioid, pleomorphic), malignant granular cell tumor, malignant Triton tumor, conventional epithelioid sarcoma (“distal-type”, classic form § and variants, any type – conventional, epithelioid, poorly differentiated), intimal sarcoma, glomangiosarcoma, extra-renal malignant rhabdoid tumor, fibroma-like, angiectoid, proximal-type epithelioid sarcoma, extraosseous osteosarcoma (any type – conventional and giant-cell rich), alveolar soft part sarcoma, clear cell sarcoma, soft tissue PEComas (monotypic epithelial angiomylipomas), Ewing’s sarcoma / primitive neuroectodermal tumor, desmoplastic small round cell tumor, high grade malignant fibrous histiocytoma group (pleomorphic malignant fibrous histiocytoma (undifferentiated pleomorphic sarcoma), osteoclastic giant cell malignant fibrous histiocytoma (undifferentiated pleomorphic sarcoma with osteoclastic giant cells), inflammatory malignant fibrous histiocytoma / undifferentiated pleomorphic sarcoma with prominent inflammation).

*Primary analogue high grade tumors also described in cutaneous site, for which the predicted prognosis should not be applied.* § Definitionally high grade sarcomas. May give rise to metastasis after a long duration follow-up, despite the absence of local recurrence.

Clinical and Pathological Parameters influencing Prognosis (“Prognostic Predictors”) 265-272

Clinicopathologic features 255-257 265 245a 271 – e.g. anatomical sites (abdominal cavity, retroperitoneum, head-neck worse than limbs, and abdominal and chest wall); tumor location (deep soft tissue worse than skin and subcutis); tumor size (the larger the worse); patient’s age (old age generally more adverse than young); clinical setting (secondary sarcomas arising in benign tumors, immunosuppression-associated tumors, virus-induced tumors, ...); others ... Grading systems and histological grades 265-271 – e.g. grade I, II, III according to the French/FNCLCC grading system, which evaluates 3 independent parameters (tumor differentiation, mitosis count, and necrosis) each scored 1 to 3: grade I → 2-3; grade II → 4-5; grade III → 6-8. Special histological types 269 – e.g. definitionally low grade tumors, definitionally high grade tumors, non-gradable tumors, tumors with not-yet-established grading parameters. Clinical stage 272 – e.g. TNM/UICC staging system: stage I (A: low grade, small, superficial, and deep; B: low grade, large, superficial); stage II (A: low grade, large, deep; B: high grade, small, superficial, and deep; C: high grade, large, superficial); stage III (high grade, large, deep), stage IV (any metastasis).

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TUMORAL, QUASITUMORAL AND PSEUDOTUMORAL LESIONS OF SOFT TISSUE - APPENDIX


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Table XXV


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Lipoleiomyosarcoma (2004;96:481-95 / III-5)

Lipomatous hemangiopericytoma (2004;96:436-63 / II-5)

Localized hypertrophic (mono)neuropathy (2005;97:92-114 / V-5A)

Low grade fibromyxoid sarcoma (2004;96:436-63 / II-11)

Mixed tumor (2005;97:343-60 / VII-1A)

Myoepithelioma (2005;97:343-60 / VII-1B)

Myofibrosarcoma (2004;96:436-63 / II-3)

Myxoinflammatory fibroblastic sarcoma (2004;96:436-63 / II-8)

Nephrogenic fibrosing dermopathy (2005;97:394-408 / VIII-5)

Neural fibrolipomatous hamartoma (2005;97:394-408 / VIII-3)

Nora’s lesion (2006;98:187-208 / IX-1)

Nuchal-type fibroma (2004;96:121-42 / I-5)

Oncocytoma (2005;97:343-60 / VII-3)

Ossifying fibromyxoid tumor (2004;96:436-63 / II-10)

Palisaded myofibroblastoma (2004;96:436-63 / II-2)

Perineuriomas - intraneural and extraneural forms (2005;97:92-114 / V-5)

Pigmented neurofibroma (2005;97:92-114 / V-2)

Phosphaturic mesenchymal tumor (2004;96:481-95 / III-1)


Plexiform fibrohistiocytic tumor (2004;96:121-42 / I-3)

Plexiform xanthomatous tumor (2005;97:141-57 / VI-7)


Proximal-type epithelioid sarcoma (2005;97:141-57 / VI-2)

Rosai-Dorfman disease of soft tissue (2005;97:141-57 / VI-6)

Rudimentary meningocele (2005;97:394-408 / VIII-3)


Soft tissue perineurioma (2005;97:92-114 / V-5B)


Superficial angiomyxoma (2004;96:436-63 / II-9)

Tophaceous pseudogout (2006;98:187-208 / IX-2)

Tumoral calcium pyrophosphate dihydrate crystals deposition disease (CPPDCDD) (2006;98:187-208 / IX-2)