Acquired elastotic hemangioma (AEH) is a recently proposed entity described by Requena et al. A uniform clinicopathological presentation allowed the authors to delineate this condition as a separate entity. At present, the original series of 6 patients represents the only source of the information on this condition.

**Clinical Findings and Gross Appearances**
All patients are elderly females, with the mean age being 64 years. Clinically, the lesion is a single angiomatosus, slowly growing, irregular plaque ranging from 2 to 5 cm in diameter that shows a strong predilection for the extensor surfaces of the forearms.

**Histological Features**
Microscopically, the condition is typified by a band-like proliferation of capillary blood vessels involving the superficial and mid dermis and arranged parallel to the epidermis (Fig. IV-1i). The vessels possess well-formed round or elongated lumens lined by a single layer of monomorphic endothelial cells (Fig. IV-1ii→IV-1iii). Cellular atypia is absent, and mitotic figures are scarce. Collagen bundles surrounding or intermingled with the neoformed capillaries typically display intense solar elastosis. The capillary proliferation is separated from the epidermis by a narrow band of spared superficial dermis. There are no hemosiderin deposits or extravasated erythrocytes.

**Immunohistochemistry**
Immunohistochemically, the capillaries have a periphreral ring of actin-positive pericytes (Fig. IV-1iv).

**Differential Diagnosis**
Histopathologic differential diagnoses of AEH include cutaneous capillary proliferations that develop during adulthood, namely Mali’s acroangiodermatitis, cherry angioma, acquired tufted hemangioma, hobnail hemangiomas and early Kaposi’s sarcoma with a predominant angiomatous pattern.

**Biological Behavior**
Acquired elastotic hemangioma is benign.
IV-2. Epithelioid hemangioendothelioma of soft tissues and skin

Epithelioid hemangioendothelioma (EHE) is a rare low grade malignant vascular tumor 1, which was first described as a distinctive entity by Weiss and Enzinger in 1982 2. EHE was so termed because its cell composition usually mimics the cell morphology of an epithelial neoplasm and the clinical course is – according to the authors who first described it – “intermediate between a hemangioma and conventional angiosarcoma” 2,3. EHE has been described in a wide variety of anatomical sites, including viscera (mostly lung and liver), where it was originally misinterpreted as an epithelial tumor being called intravascular bronchioloalveolar tumor and sclerosing cholangiocarcinoma, respectively), bone, several other various organs (brain, parotid, breast, lymph node), large body cavities, somatic soft tissues and skin. Less than one hundred cases of EHE of the somatic soft tissues 4-18 and around thirty cases of EHE of the skin 19-36 have been so far reported in detail. EHE of soft tissues and skin will be discussed here and a review of the literature of EHE in these latter locations will be offered as well.

CLINICAL FINDINGS
People of both sexes are almost equally affected and the overwhelming majority of them are adults. In the original series of 46 cases reported by Weiss and Enzinger 2, 23 out of a total of 41 patients were male and 17 female (in one case the sex was unknown) and the age ranged from 16 years to 85, with over 80% of the cases having occurred between the third and seventh decades. In the series of 30 patients described by Mentzel et al. 14 18 were female and 12 male and the age ranged from 16 to 74 years (mean 48, median 50). This lesion appears only exceptionally in children with a total of 3 cases so far on record 6,32-33, exclusive of the case reported by Vasquez et al. which occurred in a 18-year old female, probably dating back to infancy 15. Anatomical sites of involvement include the lower and upper extremities, the trunk, the head and neck area, the anogenital region. Deep soft tissues of the muscular and subfascial planes are more often affected than the superficial subcutaneous soft tissue. Cutaneous localization of EHE is less common, of which 31 total cases have been described to date 14,19-36.

EHE more often occurs as an isolated lesion, confined to the soft tissue or skin, but sometimes it occurs concomitantly with involvement of underlying bone 2,19,20,22,24 or viscera (liver and/or lung) 2,22,27,36 (multicentric or systemic form).

Soft tissue EHE usually occurs as a solitary, often painful, tumor mass. Cutaneous EHE may present in the form of an erythematous macule or elevated plaque or even as a slightly colored skin nodule, sometimes being associated with pain. Skin ulceration has been described in three cases 31,33,35. Occasionally multiple tumor lesions may also be seen (multifocal form) 2,14,29,30. Preoperative duration of symptoms varies from a few weeks to several years (up to 20 36). In some cases multicentric and multifocal forms are very difficult to differentiate from metastatic spread.

PATHOLOGICAL FEATURES
Grossly EHE of soft tissues and skin appears as a tumor mass or an indurated plaque. In around half of the cases arising in soft tissues it originates from a large or medium sized vessel (usually a vein), expanding the vessel lumen, and growing as an intravascular mass and mimicking an organizing thrombus, but being firmly attached to the vessel wall and infiltrating the surrounding soft tissue. On cut surface it is usually pale-gray, but occasionally it may show a slight reddish color, which may be indicative of its vascular nature. Tumor size ranges from few mm to several cm (0.4 to 10 cm in the series of Mentzel et al. 14); size was not given in the series of Weiss and Enzinger 2).

Microscopically in soft tissues the tumor has infiltrative margins and in half of the cases – as above said – it exhibits a striking vasculo-centric pattern of growth, infiltrating or arising from the walls of vessels. In the skin EHE may show circumscribed borders and in some cases a striking hyperplasia of the overlying epidermis may be noted, which is characterized by an anastomosing elongation of epithelial strands encircling dermal cores to such a degree that the overall picture of eccrine syringofibroadenoma is evoked. EHE is comprised of round to oval eosinophilic cells, arranged in nests or cords and embedded in a myxohyaline matrix (Fig. IV-2i). Canalized vascular differentiation is usually absent or is only seldom focally seen. Intracytoplasmic vacuolization (primitive lumen formation) is a constant feature (Fig. IV-2ii) with occasional evidence of intraluminal erythrocytes. Cytology is often quite bland and mitotic activity is usually very low (less than 1 mitosis per 10 HPF). Yet, cell pleomorphism and atypia, focal spindling of the cells, solid growth pattern, necrosis, and obvious mitotic activity, which are considered to be histological aggressive features may also be seen in some cases (see Biological Behaviour section). Inflammatory cells may be seen and a

References
1. Requena L, Kutzner H, Mentzel T. Acquired elastic hemangio-


stromal response may occur, occasionally characterized by a striking multinucleate giant cell reaction or metaplastic bone production (“ossifying variant”) and of EHE with spindle cell foci: however, the overall morphological appearances of Kaposi’s sarcoma and kaposiform hemangioendothelioma are characteristic enough to permit their correct recognition (additional- ly Kaposi’s sarcoma is HHV-8 positive). Epithelioid angiosarcoma, which portends a more hominous prognosis than EHE, is recognized on the basis of a more solid growth pattern, focal evidence of canalized vascular differentiation, geographic necrosis, more marked anaplasia and very high mitotic activity. Epithelioid sarcoma-like hemangioendothelioma is a newly recognized low grade vascular tumor, which is characterized – in addition to epithelioid features by – greater spindling of cells in a fuscicular arrangement, lack of myxohyaline stroma, lack of angiocentricity, and – immunohistochemically – consistent positivity for cytokeratins.

Cutaneous epithelioid angiomatous nodule, which is also a new benign, probably reactive, tumor-like lesion, needs to be recognized on morphological grounds. Extraskeletal chordromyxoid sarcoma and epithelioid sarcoma are both excluded mainly on the basis of their immunohistochemical profile (positivity for S-100 protein in the former, consistent positivity for cytokeratins in the latter, and negativity for endothelial markers in both).

Epithelioid neurofibroma, a recently delineated entity, is a diffuse positivity for cytokeratins, EMA and S-100 protein in all three entities, plus positivity for actin, calponin, and GFAP in the first two), histologically and ultrastructurally.

Metastatic adenocarcinoma, as indicated in the original paper on EHE, is the main condition to be differentiated. However, the histochemical staining for epithelial intracytoplasmic mucin, the expression of cytokeratins and the lack of immunoreactivity for mesenchymal and endothelial markers help to identify a metastatic carcinoma from an occult primary. Metastatic chordoma, an exceptional occurrence, can be also recognized mainly based on its characteristic immunophenotype (diffuse positivity for vimentin, EMA, cytokeratins, and S-100 protein and lack of expression of endothelial markers).

BIOLOGICAL BEHAVIOUR

Concerning EHE of the soft tissues, in the Weiss and En- zinger’s original series of 41 patients followed up for an average period of 48 months, six (13%) developed a local recurrence and 14 (31%) developed regional lymph node or lung, liver, and bone metastases. EHE “with aggressive histological features” had a more aggressive course than the ordinary forms (53% incidence of meta-
stasis versus 17%, and 31% of death versus 3%, both occurring within a shorter interval from initial diagnosis. In the series of Mentzel et al., of 24 patients followed-up for a median of 36 months, local recurrence occurred in three cases (8%) and systemic metastases in five (21%); death occurred in four (17%). On the whole it seems that up to a third of the patients develop nodal or systemic metastases and that the overall mortality is 20 per cent. Thus the overall prognosis of this tumor in soft tissue fits that of a fully malignant tumor although the prognosis is better than that of conventional angiosarcoma. Mitotic rate must be carefully evaluated (> 6M:10HPF), since this parameter seems clearly correlated with a bad prognosis. Tumor size does not seem to influence the clinical course.

Concerning with EHE of the skin it seems that EHE in this superficial location has a more favourable prognosis. Out of 17 patients reviewed by Quante et al. (eight from their own original series plus nine collected from the previous literature), two recurred locally and one developed nodal and lung metastases. However, the follow-up was too short (one month to 3.5 years in 12 patients, mean 16.4 ± sd, median 14.5) in order to derive conclusions. No cutaneous case with aggressive histological features has so far been observed.

For the time being it must be concluded that, although prognostication is possible with histologically “malignant” or “aggressive” cases, it is impossible to predict prognosis in typical cases, since ordinary EHE does have the potential to metastasize and kill the patient. Long-term studies are still needed in order to assess this important issue. Additionally in some of the cases this matter is even more difficult to assess since the criteria for discriminating between multifocality and multicentricity on one hand and local and distant metastatic spread on the other, respectively, could not be established.

References

Capillary hemangioblastoma (CH) was first described in the cerebellum by Lindau. He noted its association with retinal vascular tumors, previously described by von Hippel. These retinal tumors are histologically identical to cerebellar CH and to CH also described in visceral organs in the clinical setting of von Hippel-Lindau disease (VHL).

**IV-3. Soft tissue capillary hemangioblastomas**

CH may occur in any part of the central nervous system (CNS) (neuraxial CH). Sporadic CH occurs predominantly in the cerebellum, whereas VHL-associated CH is localized in the cerebellum, brain stem and spinal cord. Supratentorial locations of CH are rare in VHL. VHL patients often have multiple CHs at various sites; thus multiple tumors almost always indicate VHL. Rarely CH may also affect spinal nerve roots, filum terminale and cauda equina (perineuraxial CH) either in the context of VHL or sporadically. A minority of patients present with erythrocytosis, a consequence of tumoral erythropoietin production and some CHs contain foci of extramedullary hematopoiesis. All CHs outside the CNS published so far (extraneuraxial CH) occurred either in internal organs such as liver, pancreas, kidney, lungs or in the retroperitoneum or they arose in association with a peripheral nerve. Interestingly, even if there are several microscopically typical cases of CH occurring outside the CNS, including soft tissues, in the joint files of the authors of this paper, no comprehensive book on soft tissue tumors mentions CH as a tumor occurring in these anatomic compartments. All published cases of CH outside the CNS, with the exception of the retroperitoneal case, two soft tissue cases and the radial nerve case, which was not thoroughly worked up for VHL, arose in the setting of the stigmata of VHL.

**Pathological features**

Grossly a quite circumscribed hemorrhagic tumor mass of few cm in size is apparent. Histologically CH is composed of two main components: vacuolated stromal cells and rich capillary network. The cytoplasm is packed by numerous clear microvacuoles, typically impinging on the nucleus making small indentations into the nuclear outlines. The nuclei are often markedly enlarged and even variously atypical (Fig. IV-3i). In some areas the chromatin may be necrotic. In some areas the stromal cells can predominate so that the tumor in these areas can simulate a lipomatous neoplasm, especially hibernoma (Fig. IV-3ii). The vasculature consists of larger feeding and draining vessels (Fig. IV-3iii) and a rich network of small capillaries. A myxoid and pseudocystic change can focaly be seen. A small amount of hemosiderin within the stroma of CH is the rule.

**Immunohistochemistry**

Immunohistochemically the stromal cells are typically S-100 protein and NSE positive and in the cerebel-
lilar location also often GFAP positive, where this latter finding is usually interpreted as evidence of GFAP uptake by stromal cells from the surrounding brain tissues rather than a sign of a true astrocytic differentiation.

**Electron Microscopy**

Ultrastructurally the stromal cells contain electron-lucent cytoplasm containing lipid droplets (Fig. IV-3iv), small amount of rough endoplasmic reticulum and glycogen particles. No electron-dense structures typical of secretory granules can be seen in the stromal cells.

**Special Studies**

Molecular genetic studies of CH of the CNS showed that these tumors are strongly associated with the VHL gene mutations and deletions. However, no mutation of coding sequence of VHL gene was found in the only case so studied of CH of peripheral soft tissues.

**Differential Diagnosis**

CH containing areas with predominantly stromal cells is sometimes called cellular variant of CH. These cellular stromal areas, when present in CH in peripheral soft tissues, can cause difficulties in the differential diagnosis with malignant tumors, especially well differentiated liposarcoma. The atypism of the stromal cells may be, however, easily differentiated from the true lipoblasts by the uniform size of the cells as well as by the uniform size of the lipid vacuoles inside the cytoplasm.

Other well known vascular tumors of soft tissues can be easily differentiated from the CH by the lack of the stromal cells, which are virtually pathognomonic of CH. As the clear cytoplasm of stromal cells caused by numerous lipid containing vacuoles is a striking morphologic feature of CH, the distinction of CH from a metastatic conventional clear cell carcinoma of the kidney may cause difficulties. There are on record at least five cases of a conventional clear cell carcinoma metastatic to CH and the frequency of this phenomenon may be even underestimated. The immunohistochemical phenotype can easily distinguish CH (which is EMA and cytokeratin negative) from conventional renal cell carcinoma metastatic in the soft tissues.

**References**

22. Brodkey JA, Buchignani JA, O’Brian TF. Hemangioblastoma of
Dabska tumor (DT) is the eponymic term referring to “malignant endovascular papillary angioendothelioma”, which was first described in 1969. DT is an extremely unusual tumor of borderline malignancy, of which – including the 6 cases from the Dabska’s original report- 30 total cases have been described to date, exclusive of the two cases which were (previously published by other authors and) subsequently immunohistochemically studied by Manivel et al. and of four other cases mentioned by Folpe et al. in another immunohistochemical study which did not include clinical details.

In 2000 in a paper that was coauthored by Dabska herself, it is stated that, since the original description of 30 years before, knowledge in this area has expanded. As a matter of fact, although DT was initially considered to be associated with childhood, it has been subsequently demonstrated that it is not found exclusively in this age group, and although DT is primarily a tumor of superficial soft tissue, mainly involving the skin, it can also be found in deep soft tissues, bone, and visceral sites. Further, emphasis has been recently placed on its likely histogenesis from (or differentiation towards) lymphatic endothelium, probably of the “high” type, which is functionally linked with circulating lymphocytes. Therefore, the new designation of papillary intra/lymphatic angioendothelioma has been proposed and included in the new WHO classification of soft tissue tumors. Quite in contrast, the feeling has also been expressed by some authors that DT may not represent a distinct entity, since focal DT-like changes have been observed in some other vascular tumors as well.

All the above reasons prompted us to include DT in our list of updating Excerpta, focusing on those examples with frank histological lymphangiomatous features.

**CLINICAL FINDINGS**

DT may be seen in children, often as a congenital lesion, as well as in teen-agers, adults and even in elderly. Both sexes are equally affected. The skin is by far the most common location, but without predilection for any particular anatomical region. DT may occur in the head and neck, limbs and trunk. The clinical appearances are those of an intradermal nodule with or without subcutaneous involvement, often with a superficial pink to bluish blush which gives a hint to its vascular nature. Occasionally multiple nodules are seen. Tumor size ranges from 1 cm to 40 cm (case 5 of ref. 15). Deep soft tissue (periosteal) as well as intraosseous or visceral locations are also on record. DT has also not been infrequently seen in association with (or arising on) vascular malformations, hemangiomas, and mainly lymphangiomas, which may be already clinically known or may become evident on imaging or pathologic investigations. One of these cases had been clinically complicated by Kasabach-Merritt syndrome.

**PATHOLOGICAL FEATURES**

Grossly DT is described as an ill-defined, infiltrative, dermal or subcutaneous nodular mass. On occasion cystic structures may be visible to the naked eye. Histologically the distinctive architectural pattern of DT is that of several anastomosing vascular channels as well as papillary or glomerulus-like appearances. The vascular walls as well as papillations are lined by atypical endothelium with characteristic hobnail or matchstick configuration. Endothelial cells comprising the intraluminal papillations are often seen encircling acellular collagenous cores in a rosetting fashion. Mitotic figures are absent or very rare. Necrosis is not seen. Additionally a solid cellular component with epithelioid features, arranged in short non-luminized cords dissecting the stroma, may also be noted as a part of the lesion. Endothelial cells comprising these solid aggregations usually exhibit intracytoplasmic vacuolization. An intravascular and perivascular lymphocytic inflammatory infiltrate is a constant feature.

Sometimes in a DT an adjacent lymphangioma or an adjacent clustering of lymphatic vessels may be documented or in some other cases papillary or glomeroendothelial proliferations may be present as mural nodules (“in situ” component) in the lymphangiomaticous cavities, attesting to the derivation of DT in lymphangiomas. Lesions with histological features of hemangioma or non-descriptive vascular...
malformation may also be seen in the background, this
also providing evidence of the source of DT5 11 12.

IMMUNOHISTOCHEMISTRY
AND ELECTRON MICROSCOPY

Immunohistochemically, DT is usually positive for vi-
mentin, standard endothelial markers (diffuse and
strong positivity for CD31, FVIII-Rag, and Ulex Euro-
paeus-1 lectin; weak and focal for CD34), and VEG-
FR3 (vascular endothelial growth factor type 3), a new
marker of lymphatic endothelium15 20. A pericytic com-
ponent in the cell composition of the intraluminal en-
dothehial tufts is evidenced by positivity for alpha-
smooth muscle actin. The acellular collagenous cores
of the same tufts are positive for collagen IV. Cytoker-
ratins, EMA, Leu-M1, CD45, S-100 protein, and de-
smin are consistently negative.

Ultrastructurally, tumor cells show cytoplasmic inter-
mediate filaments of vimentin type, occasional Weibel-
Palade bodies, many pinocytotic vesicles, primitive in-
tercellular junctions.

DIFFERENTIAL DIAGNOSIS AND COMMENTARY

Several vascular tumors, including retiform heman-
gioendothelioma, epithelioid hemangioendothelioma,
papillary intravascular endothelial hyperplasia, glome-
ruloid hemangioni, tufted angioma, targetoid hemosi-
derotic hemangioni, conventional angiosarcoma,
epithelioid hemangioni and Kimura’s disease, reactive
angioendotheliomatosis, and the so-called malignant
angioendotheliomatosis, may enter the differential dia-
gnosis, since DT-like changes can also be focally seen
in most of these entities.

Retiform hemangioendothelioma (RH) is the main
consideration24. RH is a low grade angiosarcoma of
the skin with the following histologic characteristics:
arborizing blood vessels arranged in a rete testis-like
pattern, prominent lymphocytic infiltrate, and intrava-
sacular papillary projections with hyaline cores. On the
basis of overlapping histologic features, some authors
have even proposed the hypothesis that RH may repre-
sent the adult form of DT24 and some others include
both RH and DT under the common label of “hobnail
hemangioendothelioma”2. Further, an immunophe-
notypic relationship has also been recently document-
ted, since both entities are immunoreactive for VEG-
FR-3 15 20 and hybrid form are also on record25. How-
ever DT may be differentiated by the lack of a retiform
pattern (no retiform architecture was seen in any of the
12 cases in the series of Fanburg-Smith et al.15), the
more extensive intravascular endothelial papillations,
and the evidence – in some case – of lymphangioma or
lymphangectasia.

Epithelioid hemangioendothelioma has a different ar-
chitecture, made of cells arranged in cords and embed-
ded in a myxoid or hyaline stroma, devoid of dilated
vessels and intraluminal tufts; yet a case of metastatic
epithelioid hemangioendothelioma in a lymph node
showed an intravascular dissemination quite remini-
scent of DT (see Fig. 8 in ref. 26).

Papillary intravascular endothelial hyperplasia (Mas-
son’s reaction) is clearly a thrombosis-based reaction
occurring in the context of an occluded vein or hema-
toma or hemangioma.

Glomeruloid hemangioni has a different clinicopatho-
logic background, usually occurring in the context of
multicentric Castleman’s disease and POEMS syn-
rome, and the proliferating endothelial cells do not exhi-
bit the characteristic hobnail morphology.

Tufted angioma (angioblastoma of Nagakawa) has a
characteristic cannonball pattern of tightly packed tufts
of primitive blood capillaries, usually punctuating the
superficial dermis and lacking intravascular papillations.

Targetoid hemosiderotic hemangioni has the distincti-
ve clinical appearance of a coloured annular papula; hi-
stologically however it needs to be differentiated from
DT, since it exhibits a superficial portion comprised of
ectatic vascular spaces with papillary projections lined
by hobnailed endothelial cells. Hemosiderin dermal de-
posits are quite evident.

Conventional angiosarcoma, which can also occasion-
ally show DT-like features27, differs from DT due to the
greater cell atypism, the piling up of the endothelial
cells, the permeative pattern with collagen dissection,
and – at least for the superficial variant – the clinical
setting (occurrence on the face and scalp of elderly pa-
tients.

Epithelioid hemangioni (angiolymphoid hyperplasia
with eosinophilia) and Kimura’s disease should not be
diagnostic differential problem.

Reactive (“benign”) angioendotheliomatosis is an un-
common condition secondary to several stimuli that ex-
clusively affects the skin, characterized by a hyperpla-
sia of endothelial cells and pericytes that can result in
the formation of glomeruloid structures, occluding the
lumina of dermal capillaries.

“Malignant” angioendotheliomatosis (angiotropic B-cell
non-Hodgkin lymphoma) is easily assessed by immu-
nohistochemistry.

BIOLOGICAL BEHAVIOUR

DT is a low grade or borderline malignancy, which is
locally invasive and has the potential to metastasize. Of
the original series of 6 patients reported by Dabska1,
lymph node metastases were documented in 2 and local
infiltration of deeper structures in 4. One of the 2 cases
with lymph node metastasis subsequently died of lung
metastases. Follow-up ranging from 1 to 17 years
(mean, 9 years) showed no evidence of recurrence, me-
tastases, or residual disease in 8 of the 12 cases in the
series of Fanburg-Smith et al.15.

Local complete surgical excision with free margins is
the accepted mode of treatment.
References


IV-1. Acquired elastotic hemangioma
(Figures IV-1i to IV-1iv are courtesy of H. Kutzner, M.D., Friedrichshafen, Germany)

Fig. IV-1i. A band-like arrangement of small neoplastic vessels involving the dermis.

Fig. IV-1ii. A close-up view. Capillary-type neoplastic vessels with well-formed round and elongated lumens lined by a single layer of monomorphous endothelial cells.

Fig. IV-1iii. Note intense solar elastosis.

Fig. IV-1iv. A double staining for alpha-smooth muscle actin and CD31, highlighting the pericytic coat (in red) and the endothelial layer (in brown), respectively.

IV-2. Epithelioid hemangioendothelioma of soft tissues and skin

Fig. IV-2i. Irregular strands and groups of neoplastic cells infiltrating a densely collagenized dermis in a case of epithelioid hemangioendothelioma of the skin. A mitotic figure is barely visible in the center towards the left.

Fig. IV-2ii. Detail of the cell morphology in previous figure. Some neoplastic cells exhibit evident cytoplasmic vacuolation (unicellular vascular lumina).
IV-3. Soft tissue capillary hemangioblastoma

Fig. IV-2iii. Same case as the previous figures. The lymph node metastasis is pictured, which occurred after nine years since the initial diagnosis. A solid tumor with geographic necrosis is visible. Inset – upper left: Detail of the cell morphology showing epithelioid appearances and mitoses. Inset – bottom right: Detail focusing on a frank vasoformative area.

Fig. IV-2iv. Immunostaining for von Villebrand factor (FVIII-Rag). Neoplastic cells are intensely positive (compare with Fig. IV-2i).

Fig. IV-3i. The nuclei of the stromal cells are often markedly enlarged and even slightly atypical.

Fig. IV-3i. In some areas the stromal cells predominate so that the tumor may simulate a lipomatous neoplasm.

Fig. IV-3ii. Areas of the tumor reveal larger stag-horn shaped vessels.

Fig. IV-3iv. Ultrastructurally the stromal cells contain electron-lucent cytoplasm with lipid droplets, which cause the indentation of the nucleus.
IV-4: Dabska tumor arising on lymphangiomma

Fig. IV-4i. A lymphangectatic cavity in the upper dermis overlying a tumor mass which was present for many years in a young adult woman.

Fig. IV-4ii. A cluster of lymph vessels in the deep dermis (same case as the previous figure). A very early proliferation of endothelial cells is visible in a couple of lymphatic spaces, such as the one in the center of the field. Inset: a more evident but still early mural nodule of proliferating endothelial cells from another area of the same lesion.

Fig. IV-4iii. In the center: numerous intraluminal papillations with glomeruloid appearances. Endothelial cells show hobnail morphology. At the periphery: nests or short cords of endothelial cells infiltrating the deep dermis. This latter pattern has been rarely described in Dabska tumor (see Ref. 11 and Ref. 17).

Fig. IV-4iv. In the center: another lymphangiomatous cavity bridged by intraluminal papillary endothelial proliferations. Glomeruloid structures with collagenous stromal core, lined by hobnail endothelial cells, are apparent. At the periphery: non-luminized cords of endothelial cells infiltrating the dermis are pictured.