Low-Grade Periductal Stromal of Breast: a case report

Rosanna Nenna ¹
Cosimo Damiano Inchingolo ¹
Domenico Palmieri ²
Annalisa De Lucia ¹
Giusy Elicio ¹
Pina Miscioscia ¹

(¹) U.O.C. di Anatomia Patologica, Ospedale “L.Bonomo “ Andria - ASL BT
(²) U.O. di Chirurgia Senologica, Ospedale “ M. Dimiccoli “ Barletta - ASL BT
**Background**

Periductal stromal sarcoma (PSS) is an extremely rare neoplasm with no clinical or radiological specificity arising in the connective tissue of the breast, especially from the periductal stroma.

PSS is an entity that histologically overlaps with phylloides tumours though the main difference is the absence of leaf-like processes.

It is non-circumscribet malignant fibroepithelial tumor consisting of a sarcomatous spindle-cell proliferation localized around benign open ductal elements. Its therapeutic management is based on wide surgery with free margins.

Adjuvant therapies are not needed.
Materials and Methods

In April of the current year (2013), a 30-year-old woman presented to a breast specialist doctor with a painless lump of her left breast.

Upon clinical examination, in the equatorial external quadrant of the left breast, a small mass measuring approximately 2 cm in size, round in shape, with no signs of inflammation and not associated with axillary lymph nodes, was found.

A clinical diagnosis of fibroadenoma was offered, and subsequently, lumpectomy was performed.
Results

On gross examination the tumor was well circumscribed, nodular and well delineated and measured 1,8x1cm.

Histological examination revealed no leafy architecture, but it found a biphasic proliferation composed of epithelial and mesenchymal components.
The epithelial component corresponded to open ducts with cellular double layer, sometimes showed mild hyperplasia without atypia and foliated architecture.
These ductal units were surrounded by a spindle-cell mesenchymal proliferation which showed moderate cellular density, extensive atypia and nuclear pleomorphism, modest mitotic activity (3/10 HPF).
On immunohistochemistry, these stromal cells were positive for vimentin, CD34 and CD10, weakly positive for CD117 and progesterone receptor ……
.... negative for cytokeratins AE1/AE3, actin HHF35, desmin, estrogen receptor and S100.
Ki67 expression was very low ( < 5% )

Was made diagnosis of “low-grade periductal stromal sarcoma” subsequently confirmed by surgical pathologists of European Institute of Oncology (IEO) of Milan.
Discussion

Previously considered to be a variant of cystosarcoma with adipose metaplasia, PSS was recently recognized as a separate entity in the WHO classification system at consensus conference in Lyon in 2002. The incidence is found to be higher among pre- and postmenopausal women.

The scant literature available discusses this tumor as a biphasic breast tumor like cystosarcoma, but arising from periductal stroma, unlike phyllodes which arises from the intralobular stroma. An absence of phyllodes architecture with no distortion of the lobular unit is the most characteristic feature.
In contrast to others mammary stromal sarcomas (myofibrosarcoma) which displace normal mammary tissues, entrapping ducts and lobules peripherally, in PSS the spindle cells form cuffs around well-preserved ducts extending into lobules.

The histological features of PSS were definite by Armed Forces Institute of Pathology (AFIP) as follows:

1. a predominantly spindle-cells stromal proliferation of variable cellularity and atypia around open tubules and ducts devoid of a phyllodes pattern;
(2) one or, more often, multiple nodules separated by adipose tissue;

(3) stromal mitotic activity of $> \text{ or } = 3/10 \text{HPF}$;

(4) infiltration into surrounding mammary fibroadipose tissue.

The histologic grading depends on atypia and mitotic activity, so it ranges from being low-grade to high-grade PSS.
Conclusions:

PSS is a tumor of intermediate behavior. It may evolve into a classic phyllode tumor as well as a specific soft-tissue sarcoma. Because the number of reported cases in the literature is so small, the optimal means of managing PSS has yet to be established.

Resection with adequate margins is considered sufficient and axillary lymphadenectomy is not needed. Radiotherapy or chemotherapy have no benefit.
References:


