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LOW GRADE MYOFIBROBLASTIC SARCOMA OF THE BREAST, RARE PRESENTATION: A VERY RARE CASE REPORT WITH REVIEW OF LITERATURE

General Surgery

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ABSTRACT

Introduction: Low Grade Myofibroblastic Sarcoma of the Breast have been very rarely reported, with only about 10 cases in the worldwide literature till 2020. Myofibroblastic sarcoma has been known to arise mainly at head and neck regions, although it could be found at the extremities, trunk, and retroperitoneum. They are agressive tumors.

Case Presentation: A 39 year old female presented to surgery department with a history of swelling in the right breast. On examination- a mass of approx. 6x5 mm was present in the right upper inner quadrant of the breast. A wide local excision with axillary lymphnode dissection was done. On HPE, sections showed a mesenchymal tumour composed of cells arranged haphazardly and vaguely fascicular pattern. Immunohistologic staining results of tumour cells were diffusely positive for Vimentin and SMA immunomarkers.

Discussion: It has been a matter of considerable controversy whether or not a true myofibroblast can be neoplastic, because myofibroblasts can be found in the reactive conditions and benign neoplasms such as nodular fasciitis and fibromatosis.

Conclusion: A myofibroblastic sarcoma of breast is a rare tumor and the imaging results make it difficult to differentiate this type of lesion from other malignant masses.

KEYWORDS

Breast, Myofibroblasts, sarcoma

INTRODUCTION

Myofibroblastic sarcoma or myofibrosarcoma (MFS) is a malignant tumor which is composed of or originated from myofibroblasts.

The term myofibroblastic sarcoma has been described in several ways, as myofibrosarcoma, myofibroblasts sarcoma, myofibroblastic variant of leiomyosarcoma and fusiform cells sarcoma with myofibroblastic differentiation. Low Grade Myofibroblastic Sarcoma of the Breast have been very rarely reported, with only about 10 cases in the worldwide literature till 2020. Myofibroblastic sarcoma has been known to arise mainly at head and neck regions, although it could be found at the extremities, trunk, and retroperitoneum. They are agressive tumors, which may recur and metastasize. It's occurrence ranges from 36 to 81 years old, and predominate in females. Differential diagnosis includes benign myofibroblastic lesions, such as nodular fasciitis and fibromatosis and malignant lesions, such as leiomyosarcoma, fibrosarcoma, synovial sarcoma, angiosarcoma and rhabdomyosarcoma of fusiform cells. . The gold standard for the diagnosis is electron microscopy. However, it is possible to make the diagnosis of low-grade myofibrosarcoma based on histology and immunohistochemistry. The mammary myofibroblastic sarcomas have revealed predominantly low-grade cytomorphology rather than high-grade tumor mimicking malignant fibrous histiocytoma (MFH) in the primary lesions. We report a case of mammary myofibrosarcoma.

CASE PRESENTATION

A 39 year old female presented to surgery department with a history of swelling in the right breast since 3 months which has grown in size gradually. Patient had no history of mastalgia or fever or family history of breast cancer.

H/o menorrhagia was present since 1 year. On examination a mass of approx. 6x5mm was present in the right upper inner quadrant of the breast, irregular margins, immobile, slight puckering of the skin was present. USG of bilateral breast with axilla s/o single well defined hypoechoic mass having slightly irregular margins is present in the upper inner quadrant of the right breast of approximately 6x5mm at 2 O'clock position and 16x7mm lymphnode in right axilla (BIRADS-IV).On USG guided biopsy of right breast swelling showed tumour composed of sheets of spindle cells arranged in vague fascicular pattern, mild nuclear atypia and scattered few mitotic figure s/o low grade spindle cell neoplasm. On USG guided FNAC of axillary node showed lymphoid cells at various stages of development. A wide local excision with axillary lymphnode dissection was done which grossly revealed a ovoid nodular lesion measuring 6x5mm, and whitish, yellow, solid, homogenous, rubbery and myxoid cut surface. On HPE, sections showed a mesenchymal tumour composed of cells arranged haphazardly and vaguely fascicular pattern. The cells are spindle shaped and they exhibit mild pleomorphism. An occasional mitotic figure seen IHC with no necrosis s/o low grade myofibroblastic sarcoma with all resected margins free of tumour and lymph node showing reactive changes.

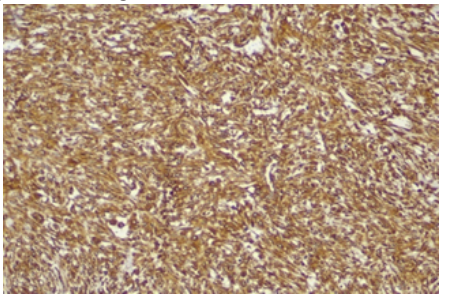


Figure 1(SMA Staining)

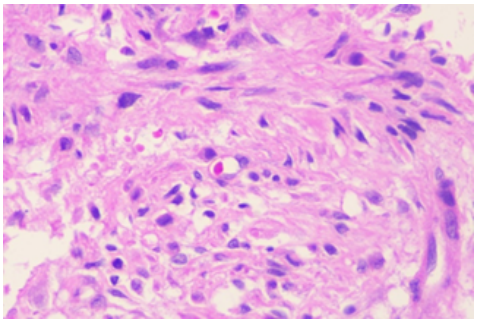
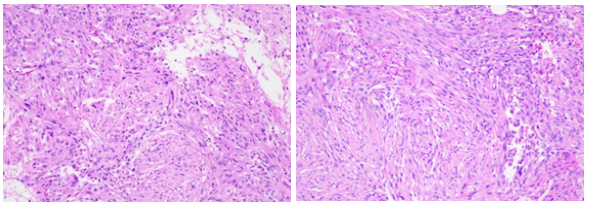


Figure 2 (A Peripheral Venule-like Vessel Reveal Intraluminally Medial Cell Proliferation)

Immunohistologic staining results of tumour cells were diffusely positive for Vimentin and SMA immunomarkers. Immunoreactivities for CD-34, Bcl2, desmin, myogenin, CK were negative. All those light microscopic findings and immunohistochemical data suggested a low grade myofibroblastic sarcoma. She got no additional chemotherapy or therapeutic radiation after wide local excision.

DISCUSSION:

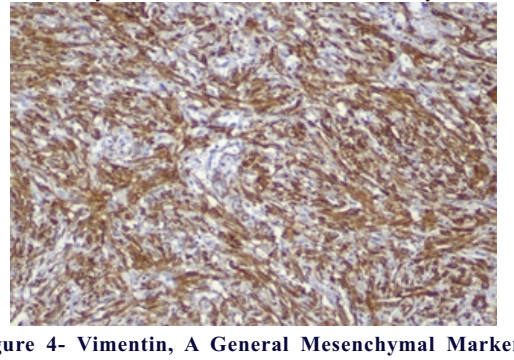
It has been a matter of considerable controversy whether or not a true myofibroblast can be neoplastic, because myofibroblasts can be found in the reactive conditions and benign neoplasms such as nodular fasciitis and fibromatosis. However, myofibroblastoma of the breast has been known as a distinctive benign mesenchymal tumor, since a study on 16 cases at 1987 showed that the myofibroblastoma cells are ultrastructurally different from fibroblasts, smooth muscle cells, or myoepithelial cells and microscopically characterized by uniformly slender and bipolar spindle cells haphazardly arranged in fascicular clusters with hyalinized fibrosis. Thereafter, malignant tumors of myofibroblasts or those with myofibroblastic differentiation have been diagnosed in the breast by the presence of more aggressive histologic findings such as frequent mitotic counts, infiltrative growth pattern, and necrosis compared to myofibroblas- tomas. They are characterized clinically by variably malignant behavior ranging from recurrence to diffuse pleuropulmonary metastasis. Myofibroblastic sarcoma or myofibrosarcoma (MFS) has been generally known to be identified best by their characteristic ultrastructural findings such as fibronectin extracellular fibrils and their fibronexus junctions, but it has been recently reported that the fibronectin can be demonstrated consistently by an immunohistochemical staining result alone. Therefore, the present case could be diagnosed as MFS showing myofibroblastic differentiation with diffuse immunoreactivity for fibronectin.



(Figure 3-lightly Fibrous Stromal Background Is Diffusely Noticed In The Highly Cellular Areas)

MFS is generally graded by two-tiered system as low and high-grades rather than low, intermediate, and high grades by mitotic count, nuclear pleomorphism, and necrosis. The former two grades of MFS share to a degree histopathologic findings of low-grade fascicular spindle cell neoplasm, whereas high-grade MFS show distinctively marked cellular pleomorphism mimicking MFH. Therefore, our case could be diagnosed cytologically as low-grade myofibrosarcoma (LG-MFS). The most representative myofibroblastic tumors of the breast are myofibroblastoma and LG-MFS. LG-MFS is a predominantly spindle cell neoplasm that could show frequent mitoses and focal necrosis, whereas benign myofibroblastic tumors including mammary-type myofibroblastoma don't reveal the two.

MFS of the breast may have a wide range of differential diagnoses such as leiomyosarcoma, fibrosarcoma, and cystosarcoma phyllodes with fibrosarcoma-like over- growth, but the most important differential diagnosis in the present case is the last one. Both lesions can show epithelial entrapment within the tumor, but there are obvious differences in morphological, immunohistochemical, and ultrastructural study results for the sarcoma cell nature. The overgrown fibrosarcoma-like lesion arising in cystosarcoma phyllodes generally show cleft-like compressed epithelial components and the fibroblastic features rather than myofibroblasts by the immunohistochemical and ultrastructural studies. Incontrast, the present tumor revealed multifocally residual linear or round ductal epithelial remnants mainly in the hyalinized collagenous stromal backgrounds, mimicking histologically ancient fibroadenoma, and immunohistochemically diffuse fibronectin reactivity in the tumor cells. With regard to the sarcoma component alone, the most important differential diagnoses may include fibrosarcoma, leiomyosarcoma, and inflammatory myofibroblastic tumor. Fibrosarcoma is composed of malignant spindle cells showing fibroblastic differentiation which is different from myofibroblasts by no immunohistochemical evidence of fibronectin, SMA, calponin. Leiomyosarcoma is also a malignant spindle cell tumor with smooth muscle features that are characterized by the presence of immunoreactivities for H-caldesmon, desmin, and occasionally keratin but no evidence of immunoreactivity for fibronectin. Thus, leiomyosarcoma and MFS can be differentiated by those immunohistochemical staining results, in spite of common immunoreactivity for SMA. Finally, inflammatory myofibroblastic tumor (IMT) can be differentiated from MFS by its microscopic findings such as diffuse inflammatory infiltrates of lymphoplasma cells, eosinophils, and histiocytes and no evidence of nuclear pleomorphism or atypical mitotic figures. Also, IMT shows a relatively frequent immunoreactivity for ALK, an important diagnostic marker for differential diagnosis with MFS. With the diffuse immunoreactivity for fibronectin, MFS can be differentiated from leiomyosarcoma and malignant fibrous histiocytoma, because the latter two use type IV collagen to connect to the extracellular matrix. Pericytes were discovered first by Charles Rouget and are generally known as mural cells or vascular smooth muscle cells because of their contractile fibers . They are quite abundant on small venules and arteries but are rather sparse on capillaries. They exhibit a number of characteristics consistent with muscle cell activity and thus can express smooth muscle actin. However, it is still not clear whether pericytes are smooth muscle cells or cells with smooth muscle cell characteristics that can turn into smooth muscle cells, which could suggest that pericytes and smooth muscle cells represent phenotypic variants of the same lineage, or even have a distinct progenitor. Therefore, pericytes share the smooth muscle actin with myofibroblast and thus have been considered a possible origin of myofibroblastic neoplasm. The present case showed some intratumoral vessels of small venule size to be irregularly thickened by mural cell proliferation of probably pericytic origin, which merged with the myofibroblastic sarcoma cells. Another putative cell origin could be the uncommitted vimentin+/CD34+ fibroblast of mammary stroma, because it is thought to be capable of multidirectional differentiation into fibroblastic, myofibroblastic, leiomyomatous, adipocytic, osseous, and cartilaginous lines . However, it is less likely candidate of tumor cell origin in this case, because the tumor cells don't' express CD34-immunoreactivity and no evidence of transition to mammary stromal cells.



(Figure 4- Vimentin, A General Mesenchymal Marker Is Immunostained Strongly In Most Tumor Cells)

CONCLUSION

A myofibroblastic sarcoma of the breast is a very rare tumor and the imaging results make it difficult to differentiate this type of lesion from other malignant masses.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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