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Case Report
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Mammary Myofibroblastoma: Case Report and Review of the Literature

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Abstract

Benign breast diseases are still considered the largest comprise of breast complaints in developing countries. They range from developmental abnormalities, inflammatory lesions, fibrocystic changes, stromal lesions, and neoplasms. Mammary Myofibroblastoma (MMF) is a rare well defined benign stromal tumor that found in both males and females. The rarity of the tumor along with its wide morphological variation challenges the accurate identification of the final histopathological diagnosis.

We present a case of a 70 years old female with 10 years history of an enlarging painless breast mass. Breast ultrasound, Mammography and MRI have demonstrated its benign nature of the large breast mass, yet the histopathology diagnosis was challenging based on the Core biopsy. It was reported as MMF. Complete surgical excision was performed with ease leaving the normal compressed breast tissue.

Introduction

MMF or myogenic stromal tumor is a rare benign breast mesenchymal tumor. It is usually found in older males and postmenopausal females as a well-circumscribed painless breast mass that seldom exceeds 3 cm. occasionally it may be incidentally discovered during surgery.

It is recognized as soft tissue tumor that arises from stromal elements giving it the ability to affect many tissues such as the breast, skin, lymph nodes and suprasellar regions of the brain [1]. When found in the breast it is labeled as "Mammary-type Myofibroblastoma".

It is composed of fascicles of spindle cells having features of myofibroblasts, spindle cells with intervening hyalinized collagenous\myxoid stroma and a variably prominent component of adipose tissue. The spindle cells characteristically express both CD34 and desmin.

In females, the diagnosis is always challenging due to the wide range of differential diagnoses. Despite the limited follow up no recurrence was reported [2].

Case Presentation

A 70-year-old Saudi female, a known case of controlled hypertension with no history of other co- morbidities presented with history of a left painless palpable breast mass for 10 years. The mass started as a small nodule which progressively increased in size with no associated symptoms apart from increased size of the affected breast and heaviness. Clinical examination revealed unremarkable general examination, local examination showed obvious asymmetry between the two breasts with no evidence of skin or nipple changes. The left breast with enlarged with a 11 cm × 12 cm painless, soft, well-circumscribed, round mass almost occupying the whole anterior breast. There were no palpable axillary nodes. The right breast was within normal limits.

The patient underwent series of specific imaging investigations which included, Ultrasound, Mammogram, contrast enhanced mammogram and breast MRI followed by core biopsy.

Ultrasound was reported an avascular heterogeneous hypoechoic mass occupying most the left breast (Figure 1).

Diagnostic mammogram showed a unifocal large well-defined macro-lobulated dense mass occupying predominantly the upper most of the anterior and mid breast that is surrounded by rim of lucency. No associated suspicious microcalcifications or architectural distortion or skin thickening. No enlarged axillary lymph nodes were noted (Figure 2).

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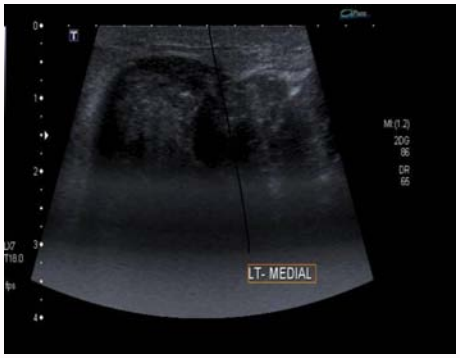

Figure 1: Ultrasound was reported an avascular heterogeneous hypoechoic mass occupying most the left breast.

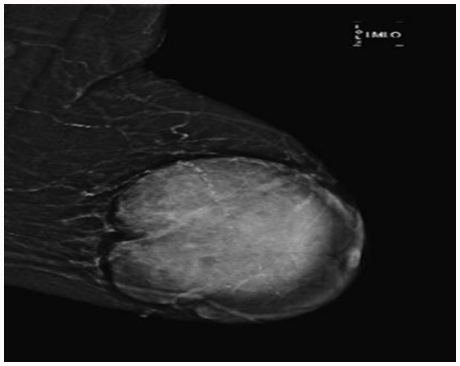

Figure 2: Mammogram showed a unifocal large well-defined mass.

MRI also confirmed the benign nature of the large 11 cm × 9 cm × 10.7 cm, well-circumscribed lobulated mass. Based on the Radiological findings it was collectively reported as BIRAD IV.

Core biopsy showed the spindle cells infiltrating the dense fibrous tissue in both linear and curvilinear arrays. Nor mitosis or necrosis was identified. Immunostaining reported Positive estrogen and progesterone receptors, and HER2 negative with Ki 67 5% (Figure 3a-3d).

Surgical excision of the mass of the mass was performed. Intraoperative findings demonstrated a well capsulated soft tissue mass with large tortuous blood supply running along the peripheral surface of the lesion. The lesion shelled from the compressed non-infiltrated breast tissue with ease.

The diagnosis was confirmed with further staining which showed desmin, calponin, CD34, (Figure 4a,4b).

The patient underwent smooth postoperative recovery and was discharged with regular outpatient follow up with no recorded complications or recurrence in six months follow up.

Discussion

MMF is a rare entity that was first described as a distinct benign mesenchymal breast lesion by Wargotz et al. [1]. It gained the term mammary type-Myofibroblastoma after challenging diagnosis based on diligent staining to exclude other possible differential diagnoses, and thus was recognized [3].

MMF is reported in both males and post-menopausal females with an average age of 63 years. The average size at diagnosis is around 3 cm; however, larger sizes have also been reported. It has

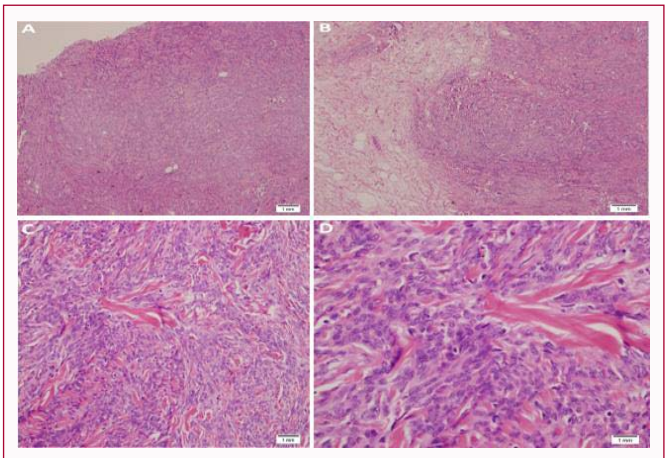

Figure 3a-3d: H&E slides (**a:** × 4 magnification, **b:** × 10 magnification **c:** × 20 magnification, **d:** × 40) showing a well-demarcated cellular neoplasm with pushing borders. It is composed of bland spindle cells, eosinophilic cytoplasm running in fascicles separated by thick eosinophilic collagen bundles; cells have an ovoid to elongated nuclei with mild nuclear polymorphism and conspicuous nucleoli.

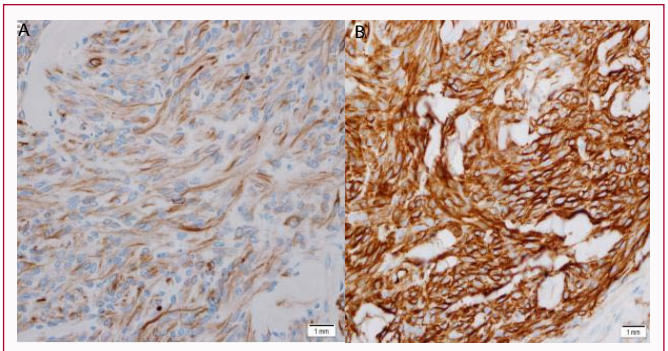

Figure 4: Immunohistochemical profile of the neoplastic cells demonstrating **a:** Desmin and **b:** CD34.

non-specific clinical and radiological characteristics and usually presents as a single painless, non-tender mass that has the tendency to increase in size [1,3]. Diagnostic imaging have reveals it's well demarcated nature however a variety of differential diagnoses of other well circumscribed lesions such as phyllodes tumor, Pseudo Angiomatous Stromal Hyperplasia (PASH) or atypical fibroadenoma should be differentiated [4]. The point of differentiation is the total absence of epithelial elements in MMF.

In females, the wider range of differential diagnoses usually hinders the quick and accurate identification of this unusual entity [5].

Histologically the classical picture is of a well demarcated but not encapsulated tumor borders composed of bland spindle cells running in fascicles separated by thick hyalinized collagen bundles. The neoplastic cells have slightly eosinophilic cytoplasm and oval nuclei with conspicuous small nucleoli. Typically, the tumor has low mitotic activity and there is no evidence of necrosis, atypical mitosis or entrapment of mammary epithelial tissue [1-3].

Nevertheless, more histological variations were also reported. Detailed contemporary morphological review revealed variants of MMF including classic, myxoid, lipomatous, epithelioid and deciduoid, along with the differential diagnoses of each type, and emphasizing the importance of recognizing MMF with all its variants [4].

MMF may mimic many stromal lesions rendering the diagnosis challenging. Reported cases of MMF imitating invasive lobular carcinoma, Schwannoma and metaplastic carcinoma by H&E stain. This confusion commonly resolved by immunohistochemical profile [5,6]. Immunohistochemistry may show tumor cells are positive for CD34 and desmin in approximately 89% and 91% of the cases; respectively. Tumor has been reported to be positive for BCL-2, CD99 and Smooth Muscle Actin (SMA). Other immunohistochemical markers that may aid in the diagnosis include Pan-cytokeratin and S100; both are typically negative in the neoplastic cells [2].

The well-circumscribed nature of the lesion coupled with the low proliferation index and absence of marked nuclear pleomorphism favors the benign diagnosis.

Pseudo Angiomatous Stromal Hyperplasia (PASH) can seldom be confused with MMF since a well-defined mass rarely occurs [7].

It should be emphasized that MMF may occur at different sites. Clinicians should keep the diagnosis in mind when they are confronted with lesions of the prostate, liver, vulva and the groin/inguinal region [8,9].

Multimodality imaging techniques aid in the diagnosis. Mammary MMF is the most disclosed lesion due the breast screening programs, the availability of imaging modalities and the ease of accessibility to the affected tissue [10].

This presented case revealed the benign nature of the disease from the long history and clinical presentation. The lack of pain and other local symptoms precluded the early presentation. Large size breast lesions may equally be reported in male patients [10,11].

Despite the benign nature of the lesion, the recognition and surgical excision should be performed. Pressure on vital structures such as the brain may cause unwarranted outcomes. In the Breast local surgical excision is the treatment of choice.

Conclusion

MMF is a challenging histological diagnosis. Surgical excision is the treatment of choice. No Adjuvant treatment is required.

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