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A case report

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Breast myofibroblastoma in a young woman. A case report

Myofibroblastoma (MFB) is an uncommon benign mesenchymal tumor that may arise in several organs and tissue. Although most of reported cases were located in the breast, it is extremely rare, representing less than 1% of breast tumor. MFB has predominantly seen in elderly men, but some cases have been described in menopausal women. This lesion is a stromal tumor which has many morphologic variants including cellular, collagenized, epithelioid, palisaded, lipomatous, hemangiopericytoma-like, and infiltrant features. Even if its incidence has recently increased due to the mammary screening, only few cases have been reported in Literature and even less in young women. Physical examination discloses a solitary, unilateral, painless, freely movable, usual firm in consistency, non-tender nodule. Imaging investigations usually are not specific to establish the right diagnosis. Furthermore, findings from Fine-Needle Aspiration (FNA) may be confusing and nonspecific, making diagnosis of MFB possible only after surgical operation. Not evidence of malignant transformation, recurrence or distant metastasis after a follow-up period of 15 years have been reported in Literature when resection margins are free. Hereby the authors describe a rare case of breast MFB in a young woman.

KEY WORDS: Breast neoplasm, Myofibroblastoma, Premenopausal woman

Introduction

Myofibroblastoma (MFB) of the breast is a rare benign mesenchymal tumor of the mammary stroma predominantly seen in elderly men even though some cases have been described in menopausal women [1]. MFB was described for the first time by Wargotz et al. [2] in 1987. It has its own pathological identity although it shares several similarities with stromal tumors arising from the lower female genital tract and other tumors classified as spindle cell neoplasm as solitary fibrous tumor, leiomyoma and benign fibrous histiocytoma [3].

Physical examination discloses a solitary, unilateral, painless, freely movable, usual firm in consistency, non tender nodule that has been growing slowly during the course of several months to years [2, 3]. More rarely, patients complain of massive enlargement of breast due to a giant tumor [4].

The X-ray mammography, Ultrasound and MRI findings of MFB usually are not specific [3,5-7] to establish the right diagnosis. Furthermore, findings from Fine Needle Aspiration (FNA) may be confusing and nonspecific [6], making diagnosis of MFB possible only after surgical operation.

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Case report

A young woman presented to our hospital with a palpable mass of her right breast arised at the age of 39 and it grew slowly in the last two years. There was no history of pain or fever associated with the swelling and not family history of breast cancer. She was in general a healty woman. Physical examination just revealed a relatively movable mass in the upper inner quadrant of the right breast. The left breast was normal and there was no palpable axillary lymphadenophaty. Based on clinical and imaging findings of the breast lump, it was thought to be a solitary fibrous tumor of the breast and this is the reason why she did not undergo surgery previously.
Routine baseline pre operatory investigations including chest X-ray, laboratory tests and ECG were performed resulting in normal limits. Mammography (Fig. 1) and Ultrasound scan of the right breast, showed a 2 x 2 cm circumscribed solid nodular mass without abnormal calcifications. No sonographically guided FNA was performed. The patient underwent a lumpectomy under local anesthesia with extemporaneous histological diagnosis of solitary fibrous tumor. The postoperative period was uneventful. Gross examination revealed a 1.8 cm, well demarcated nodular tumor with white-gray cut surface. There was a free resection margin measuring 5-10 mm. However definitive histological examination revealed a well definite and pseudo-encapsulated tumor surrounded by mammary tissue with mesenchymal appearance. Tumor cells showed oval nuclei without atypia and with very scant mitotic activity. Cells were arranged in ill-defined fascicles, haphazardly intermingled with coarse short bundles of collagen. There were no areas of necrosis, hemorrhage or metaplastic changes.

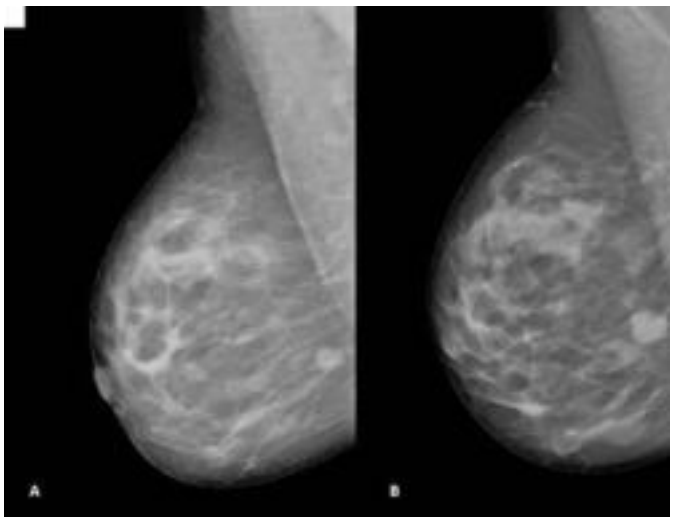

Fig. 1: Mammography depicting a discrete nodule in the upper inner quadrant of the right breast increased in volume in the last two years. A Mammographyc image in 2010; B The same image in 2012.

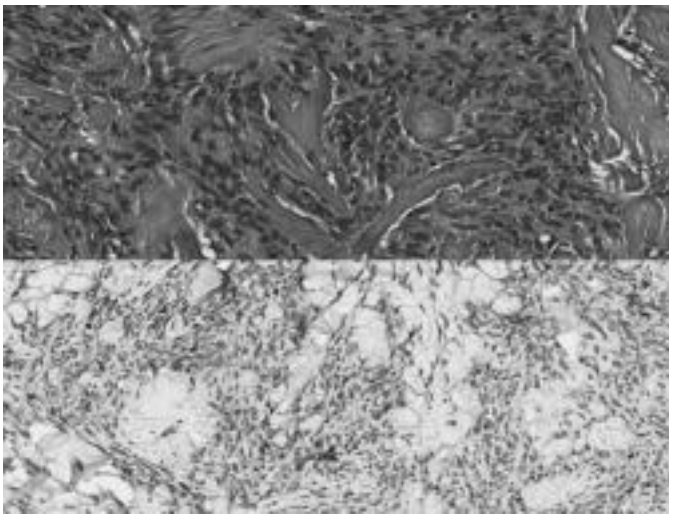

Fig. 2: Pathologic examination. The lesion show bundles of slender, uniform, spindle shaped cells typically arranged in clusters and bands of hyalinized collagen distributed throughout the tumor, Haematoxylin and Eosin, 400x (upper) and Desmin 300x (lower).

Immunochemistry revealed a positive reaction of tumor cells for desmin while reaction for actin was negative (Fig. 2). The proliferative fraction of tumor cells, detected with Ki-67, was 3%. Present findings were consistent with a diagnosis of MFB.

Discussion

MFB is an uncommon benign mesenchymal tumor that may arise in several organs and tissue, including soft tissue of retroperitoneum or inguinal area, abdominal wall, meninges, suprasellar area, vagina and vulva [8]. Although most of reported cases were located in the breast, it is extremely rare, representing less than 1% of breast tumor. Since Wargotz's description in 1987, little more of 70 cases of breast MFB have been reported in Literature [8]. It has been originally described as a tumor typical occurring in the breast of adult males with a median age of 64 years [2,8]. After about 30 female cases (range 40-87 years) have been documented in Literature it has been understood that MFB can occur in both sexes [1].
Furthermore it has been described only one case in an adolescent boy [3] and recently only one in an infant male [9]. MFB has been documented also in irradiated breast for intraductal carcinoma [10] and it has been reported only one case, in a 25-year-old woman as bilateral MFB [8].
Breast soft tissue neoplasms composed by myofibroblasts have been classified as MFB. It exhibits several morphological features characterized by fascicles of spindle cells with large hyalinized collagenous stroma, without necrotic or hemorrhagic areas, and having a surrounding pseudocapsule composed of compressed breast tissue [2].

The histological variants of this tumor include cellular [11], collagenized, epithelioid, palisaded, lipomatous [12], hemangiopericytoma-like, and infiltrant features.
Association of cartilaginous or smooth muscle areas and giant cells components were also reported [6]. Sometimes, myofibroblastoma may arise in a mammary hamartoma and may present interspersed epithelial structures, which may increase the difficulty of diagnosis [8]. Furthermore there are cases with infiltrating patterns and calcifications seen on mammography[13], or collision tumors [14]. Finally Gurzu et al.[8], reported classical cellular variant associated with a rich reticulinic network and scanty collagen bands, which may lead to an incorrect diagnosis.
The imaging findings of MFB are not specific [3,5-7]. The mammographic findings usually consist of a well-circumscribed round to oval dense and non-calcified mass. On sonography, MFB shows a homogeneously hypoechoic well-circumscribed solid mass [3,5-7] that resembles the one of fibroadenoma. Usually MRI of MFB show a homogeneous enhancing mass with internal septations [7]. FNA cytologic examination is nonspecific, although it should permit differentiation between benign and malignant proliferation [15].
Currently in Literature just few cases of premenopausal breast MFB have been described and nobody knows exactly the reason why. It is crucial to understand the molecular mechanism, controlling growth and proliferation of myofibroblasts. It is believed that this mechanism is somewhat related to autocrine and paracrine secretion of cytokines, which transform growth factors in a particular way (TGFβ). On the other hand it has also been demonstrated that other specific factors such as tumor necrosis factor (TNF) and fibroblastic peptide-trophic growth factors are generally present in these tumors[1]. Further studies are necessary to elucidate the molecular mechanism of controlling growth and proliferation of myofibroblasts.

Conclusions

MFB can occur in a young woman. It is a well encapsulated tumor with a good cleavage plane, which usually allows easy surgical local excision. Imaging and FNA are not specific to establish the right diagnosis and surgery is the only recommended diagnostic and curative treatment. Not evidence of malignant transformation, recurrence or distant metastasis after a follow-up period of 15 years have been reported in Literature when resection margins are free[1]. However, a minimum of 24 months' follow-up is desirable.

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Riassunto

Il Miofibroblastoma (MFB) è un raro tumore mesenchimale benigno che può insorgere in molti organi o tessuti. Sebbene molti casi riportati in Letteratura sono descritti a livello mammario, esso è estremamente raro, rappresentando meno del 1% di tutti i tumori della ghiandola mammaria. Il MFB è prevalentemente riscontrato negli uomini adulti, tuttavia alcuni casi sono anche stati descritti in donne in età menopausale. Il MFB è annoverato tra i tumori di origine stromale e mostra diverse varianti morfologiche includendo quella a tipo cellulare, a fibre di collagene, epitelioide, a palizzata, lipomatosa, tipo emangiopericitoma e in ultimo quella infiltrante. Anche se la sua incidenza è recentemente aumentata a causa dello screening mammografico, solo pochi casi sono stati descritti in Letteratura e ancora meno in donne in giovane età. L'esame obiettivo mostra un singolo nodulo, unilaterale, non doloroso, mobile sui piani sottostanti e di consistenza dura. Le indagini radiologiche non sono specifiche per giungere ad una corretta diagnosi preoperatoria. Inoltre anche i reperti ottenuti mediante aspirazione con ago sottile potrebbero generare confusione rendendo di fatto possibile la diagnosi di MFB soltanto all'esame istologico definitivo analizzando il pezzo operatorio asportato chirurgicamente. Nessuna evidenza di trasformazione maligna, recidiva o metastasi a distanza dopo un periodo di follow-up di 15 anni sono state descritte in Letteratura quando i margini di escissione sono risultati liberi da malattia. Di seguito gli Autori descrivono un raro caso di MFB della mammella in una giovane donna.

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