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Fibromatosis of the Male Breast: Role of Dynamic MRI

Case Report

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Abstract

Fibromatosis of the breast is a non-metastasizing benign, but locally invasive stromal tumor, accounting for less than 0.2% of all primary breast lesions and is formed by proliferation of fibroblastic and myofibroblastic cells. Mammary fibromatosis in men is extremely rare and may mimic primary breast malignancy. We report a case of breast fibromatosis in a 52-year old male, where magnetic resonance imaging (MRI) was performed, which was showing some salient features by which it can be distinguished from breast carcinoma and confirmed on histopathology and Immunohistochemical (IHC) examination..

Keywords: Fibromatosis; Mammography; Ultrasound; Magnetic Resonance Imaging

Case Report

We report a case of a 52-year old male presented with the chief complaint of a slowly progressive painless left breast lump for one year. Clinical examination revealed a firm painless swelling in the upper outer quadrant of the left breast with no palpable lymph nodes and nipple retraction. Family history was non-contributory and there was no history of trauma or surgery.

On mammography (Alpha RT-GE), an irregular shape, non-calcified high-density mass with spiculated margins seen in the left breast, in the upper outer quadrant, posterior depth and classified as BIRADS 4C lesion. At the ultrasound (Samsung RS 80A), the mass was seen as an irregular hypoechoic mass with spiculated margins and posterior acoustic shadowing suggestive of malignant lesion BIRADS 4C. There was no involvement of pectoral muscle and the axilla was normal.

MRI (GE Signa Excite, using a dedicated 8 channel breast coil) was performed to delineate tumor extent and preoperative planning. It showed an irregular shaped mass with spiculated margins in the left breast measuring 3.5x 1.7x 2.5cm appearing isointense to muscle on T1 Weighted image (Figure 1), hyperintense on T2, and T2 FAT SAT images (Figure 2). The mass was seen separate from the pectoral muscle and fascia. Pre contrast image (Figure 3). Dynamic postcontrast scans reveal progressive homogenous enhancement (early, Figure 4) and consistent with type I kinetic curve (delayed image, Figure 5). No evidence of lymphadenopathy was noticed. On Diffusion-weighted imaging (DWI), restriction with an ADC value of 1.47e -09 was seen. Diffuse glandular type of gynecomastia of left breast was noticed. The right breast was normal. Based on MRI possibility of BIRADS 4C lesion was given by looking into its morphology. Sonographic guided core biopsy of the mass was performed by 14 G trucut needle, demonstrating a low-grade myofibroblastic proliferation along with spindle cells consistent with breast fibromatosis. The patient underwent a simple mastectomy. On gross examination 4.5 x 3x2.5 cm

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tumors was identified in the upper outer quadrant. Fibrous spicules were seen radiating from the tumor. Histopathological sections studies showed bundles of fibro- collagenous tissue consisting of benign spindle cells, with sparse mitosis and no evidence of necrosis (Figure 6). Overlying skin, nipple-areola, and deep surgical margins were free of infiltration.

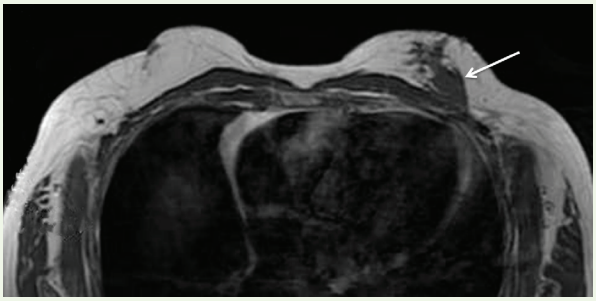

Figure 1: T1 Weighted MR image showing an irregular shape, spiculated margin, isointense mass in left breast in the posterior third region adjacent to the pectoral muscle.

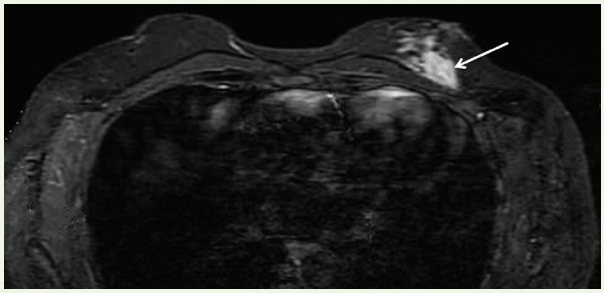

Figure 2: T2 FATSAT image showing hyperintense mass.

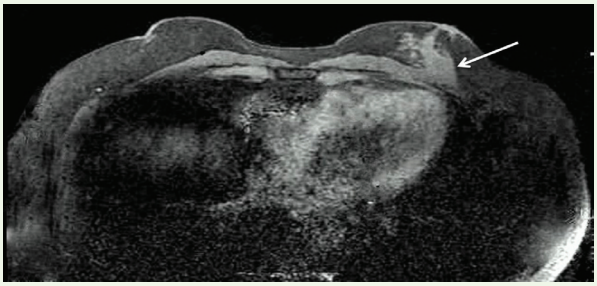

Figure 3: Pre-contrast image.

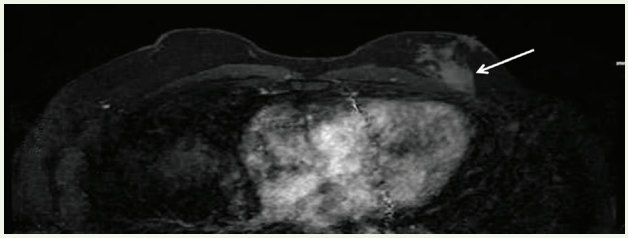

Figure 4: Dynamic postcontrast scans reveal progressive enhancement (early).

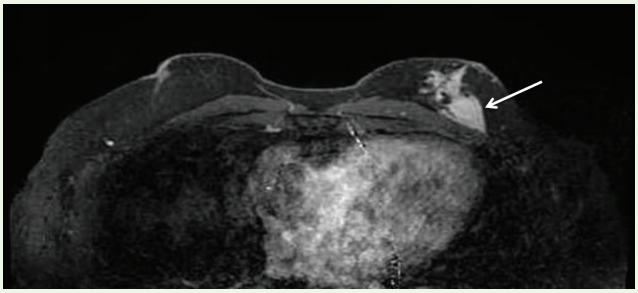

Figure 5: Delayed post-contrast image.

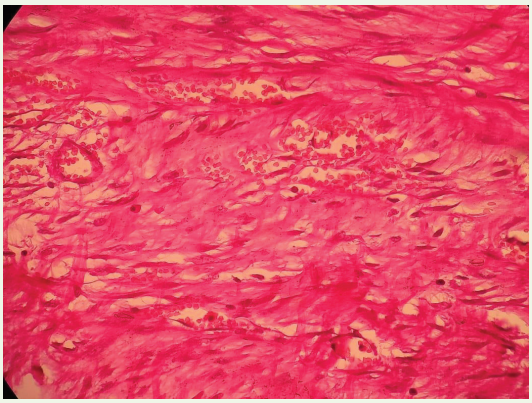

Figure 6: Histopathologic findings showing, bundles of long sweeping and intersecting spindle cells with collagen tissues.

Immunohistochemistry was negative for pan-cytokeratin, desmin, S-100, CD 34. Vimentin and smooth muscle actin were positive with a Ki67 index of less than 1%. Estrogen (ER) and progesterone (PR) receptors were absent. Based on histopathology and IHC a diagnosis of breast fibromatosis was made.

Discussion

Mammary fibromatosis is a rare entity, accounting for less than 0.2% of all breast tumors. The patient's age ranges from 13 to 83 years with the vast majority of cases occurring in women. Very few cases have been reported in men [1].

The etiology of this lesion is not well understood, but an association with Gardner's syndrome has been reported. Additional associations include familial multicentric fibromatosis, silicone and saline breast implants, incidental, and surgical trauma. Fibromatosis may arise from the pectoralis muscle or fascia or the mammary tissue. Clinically, desmoid tumors of the breast present as a firm painless, movable mass, with or without skin retraction and dimpling. Nipple retraction is seen if the tumor is close to the nipple. Nipple discharge and palpable lymphadenopathy are not associated with breast fibromatosis [2].

Multiple imaging modalities have been used to characterize breast fibromatosis, however, the final diagnosis is based on Histopathological findings. On mammography desmoid tumors present as high-density non-calcified lesions, with irregular shape

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and spiculated margins mimicking breast carcinoma. On Ultrasound, breast fibromatosis frequently appears as a poorly defined, hypoechoic mass with a thick echogenic rim and a posterior attenuation. It is not associated with adenopathies. Ultrasound Elastography was not found to be useful in discriminating between mammary fibromatosis and malignant tumors in the breast, because the composition of mammary fibromatosis lesion makes it stiffer than normal breast tissue and may lead to a false diagnosis of malignant tumor based on the elastographic result [3].

Breast fibromatosis on MRI appears as irregular, hypointense to isointense on T1 weighted images, and hyperintense on T2-Weighted images. The lesion usually shows progressive or plateau type of enhancement on the post-contrast scan as compared to rapid enhancement of the malignant lesion. Our case also had T1 isointense and T2 hyperintense irregular mass with progressive contrast enhancement. MRI is useful to show chest wall involvement, which is important for surgical planning [4,5].

Cytologic examination by fine-needle aspiration is usually not diagnostic. Definite diagnosis is made by diagnostic surgical biopsy. Histologically the lesion is composed of bundles of long sweeping and intersecting spindle cells with collagen deposition. Mitotic figures are rare. Differentials include scar or keloid, nodular fasciitis, schwannoma, leiomyoma, solitary fibrous tumor, spindle cell lipoma, myofibroblastoma, myoepithelioma, low-grade fibromyxoid sarcoma, and low-grade fibrosarcoma. The presence of spindle cells admixed with epithelial cells should raise the possibility of fibroadenoma, phyllodes tumor, or metaplastic spindle cell carcinoma. In fibromatosis, myoepithelial markers are absent [6,7].

Immunohistochemically, fibromatosis exhibits positivity for smooth muscle actin and vimentin and negativity for cytokeratin, estrogen, progesterone, and androgen receptors. Desmin is rarely positive, whereas S100 and CD 34 are usually negative [7,8].

Fibromatosis in the breast differs from fibromatosis arising in another part of the body due to its hormone receptor profile. Although 30 % of extramammary fibromatosis is positive for ERs, only one of the previously reported cases of mammary fibromatosis expressed hormonal receptors. A positive reaction of ER and PR in the spindle cell neoplasm of the breast might help exclude fibromatosis from its differential diagnosis.

Fibromatosis is a benign entity without metastatic potential but carries a significant risk for local recurrence. The breast is an unusual location for the development of this tumor, with relatively few cases reported in the literature. Although it does not metastasize, it is frequently locally aggressive and is proven to recur (up to 35%) even after complete surgical excision with clear margins. Skin retraction is caused by fibrous tissue contraction as compared to the desmoplastic reaction which is similar to tethering associated with malignancy. The clinical presentation and the radiological appearance of breast fibromatosis are highly suspicious for breast carcinoma. The tumor is best differentiated by histopathology and immune histochemistry study.

Treatment consists of wide local excision with clear margins, other options include radiation therapy and chemotherapy in patients who are not surgical candidates [9,10].

To conclude male breast fibromatosis is a rare, locally aggressive, benign breast tumor that mimics breast cancer on mammography and ultrasound but MRI can help in its diagnosis, by showing benign nature, which can be confirmed with an appropriate histopathology and immunohistochemistry study.

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