

Case Report

Extra abdominal mammary fibromatosis (Desmoid tumor) of breast in an elderly female

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ABSTRACT

The extra-abdominal form of desmoid tumor is very rare and the incidence of mammary desmoid tumor is <0.2% of primary breast neoplasm. We report a case of mammary fibromatosis in an elderly female in which no associated risk factor was discernible. After simple mastectomy as per the patient's wish, in view of posterior margin positivity, adjuvant radiotherapy was given. The patient has remained recurrence free after one year of follow up.

Keywords: Desmoid tumor, Mammary fibromatosis, Extra abdominal, Adjuvant radiotherapy

INTRODUCTION

Desmoid tumors occur most frequently in the aponeurosis of the rectus abdominis muscle of multipara women and constitute 0.3% of all solid tumors.^{1,2} The extra-abdominal form of desmoids tumor is very rare and the incidence of mammary desmoid tumour is <0.2% of primary breast neoplasm.^{3,4} The origin of mammary desmoid may be from the parenchymal tissue of the breast or as an extension of a lesion arising from the muscles of the chest wall.³ An association of such tumor has been reported with Gardner's syndrome, silicone breast implants and surgical trauma.⁴ We report a case of mammary fibromatosis in an elderly female in which no associated risk factor was discernible.

CASE REPORT

A 60-year-old Asian Indian woman presented to our hospital with a three months history of right breast pain and a lump which she noticed while bathing. The patient was the mother of five children whom she had breast fed for about two years each. The patient had menarche at the

age of about 14 years and menopause at the age of 47 years with normal menstrual history. There was no significant medical or surgical history. On examination, a 2 × 2 cm lump was palpable in the right inframammary region with mild tenderness on deep palpation. The lump was firm in consistency, smooth surface, ill-defined margins with normal overlying skin. The mobility of the lump was slightly restricted but overlying skin could be pinched up easily. No axillary or supraclavicular nodes were palpable. Bilateral mammograms showed a dense background with an area of malignant appearing spiculated mass in the lower inner quadrant of her right breast. Ultrasound showed a lobulated, heterogeneously hypoechoic lesion measuring 25 mm × 22 mm × 11 mm in the lower inner quadrant of the right breast. A core biopsy was then performed which showed small clusters and discretely lying cells in a lipoproteinaceous background suggestive of a benign spindle cell lesion keeping with the diagnosis of fibromatosis of the breast. Immunohistochemistry revealed negative staining for AE1/3, MNF, S100, CD34 while it was positive for SMA (smooth muscle actin), desmin and vimentin. The specimen was negative for estrogen and progesterone receptors. Contrast enhanced computed tomography of

the abdomen and pelvis did not reveal any additional finding.

Due to complaints of breast pain and discomfort and patient's will for complete removal of diseased breast, simple mastectomy was performed with intraoperative finding of infiltration of the pectoralis major muscle which was also excised. Her post-operative recovery was uneventful. The histopathology of the specimen confirmed the findings of the core biopsy. The lesion was demonstrated to be a fibromatosis with no cellular atypia, all margins except the posterior margin were clear. The posterior margin showed infiltration. After discussion of the case in the multi-disciplinary tumor board, the patient was given radiotherapy to the right chest wall at a dose of 50Gy/25 fractions/2Gy per fraction over a period of 5 weeks. The patient is currently on follow up and has remained disease free for 1 year.

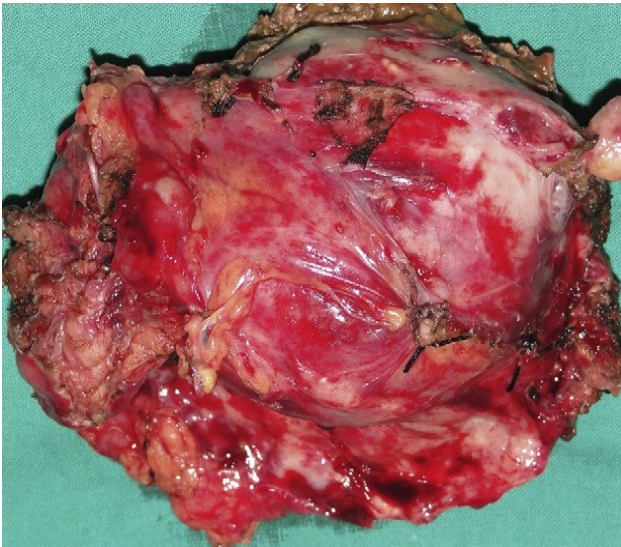


Figure 1: Resected surgical specimen of the desmoid tumor of the breast.

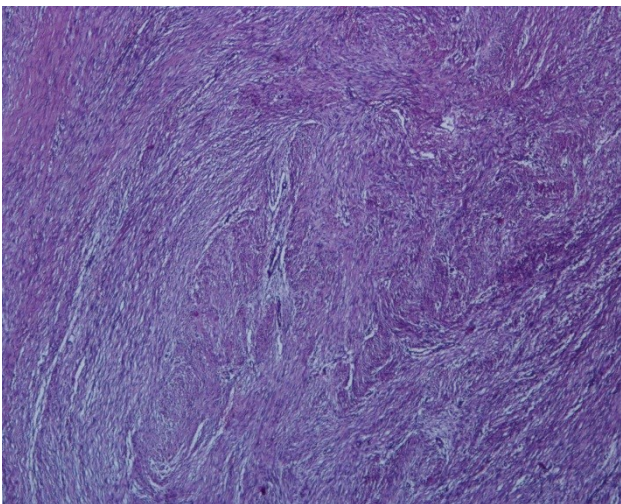


Figure 2: Photomicrograph showing H&E staining (4x) showing spindle cells.

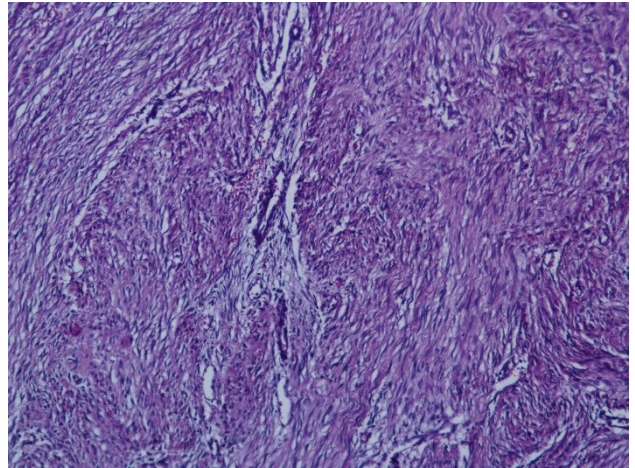


Figure 3: Photomicrograph showing H&E staining (10x) showing bland-looking spindle cells organized into long sweeping and intersecting fascicles.

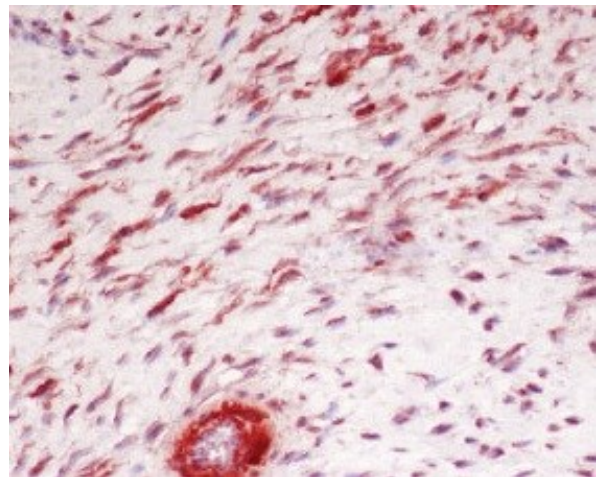


Figure 4: Photomicrograph showing positive staining for vimentin in the resected tumor tissue.

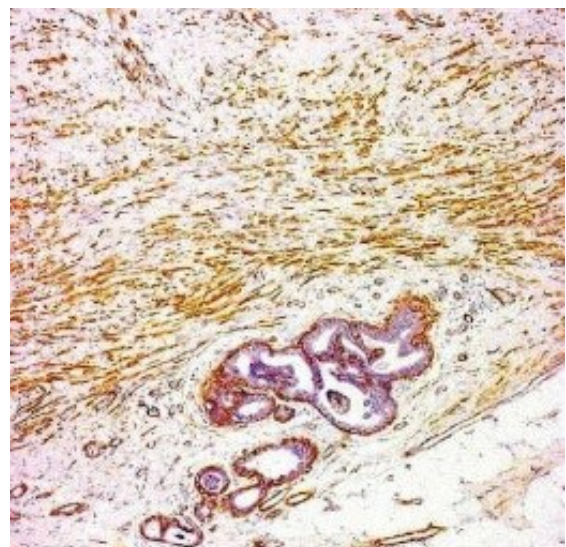


Figure 5: Photomicrograph showing positive staining with α -smooth muscle actin.

DISCUSSION

Extra abdominal mammary fibromatosis is a very rare tumor with most of the cases reported in young and fertile women with a few also reported in men.⁶ Hormonal milieu, as is the case during pregnancy, is thought of as a noteworthy predilection for developing desmoid tumor.⁷ The patient in our report is an elderly female with no such risk factors. Besides, other known risk factors, including previous surgical trauma, breast implant and Gardner's syndrome all were absent in this case.

Clinically, desmoid tumors of the breast are movable, firm masses. Skin retraction or dimpling may be present. Lesions close to the nipple may present with nipple retraction. Such skin and nipple changes were not present in our case probably due to deeper location of the lesion. On mammography, desmoid tumors are often irregularly shaped, non-calcified, high-density masses with speculated margins that mimic breast cancer as in our case.⁸ Ultrasound findings include poorly marginated, hypo echoic masses with a thick echogenic rim and posterior attenuation.⁹ On pathological examination of the specimen, desmoid tumor is usually a non-encapsulated well-differentiated fibroblastic lesion composed of relatively uniform fibroblasts and collagen and forming a firm, solitary, or multi nodular mass with an infiltrative growth pattern.¹⁰ The hallmark of a desmoid tumor is the presence of bland-looking spindle cells organized into long sweeping and intersecting fascicles. The overall cellularity is low to moderate, and there is no significant mitotic activity.¹¹ The histopathologic differential diagnosis can vary from benign reactive lesions such as a hyperproliferative scar, to a more sinister fibrosarcoma.¹¹ The distinction between a desmoid tumor and fibrosarcoma is particularly important, as the latter has the capacity for distant metastasis. Though it lacks metastatic potential, fibromatosis can grow aggressively causing local infiltration.¹² The failure to be acquainted with this process as a restricted entity within the breast and the usual temptation to stamp it as scar tissue from a previous breast biopsy or trauma may ultimately lead to local recurrence within the breast if inadequately treated.⁵ This failure can ultimately result in the need for more radical resections to obtain clearance of the surgical margins leading to poor cosmetic results.^{13,14} However, in our case, the patient did not have any history of trauma or surgery.

Because of rarity of fibromatosis of the breast, no definite guidelines have been established for the exact management of this tumor. Obtaining negative surgical margins in most patients has generally been associated with reduced possibility of local recurrence.¹² Complete wide local excision with about 1cm margin is the first-line treatment of choice. Axillary dissection is usually not performed. However, recurrence is relatively common usually occurring within 3 years of excision requiring radical surgery. A recurrence rate ranging from 21 to 57% has been noted with surgical treatment alone.^{1,3,4,15}

Margin status, large tumour size and young age have been associated with a higher incidence of recurrence.^{3,15} Postoperative radiation therapy can improve the 10-year recurrence-free survival rate.¹⁶ The role of adjuvant therapy after surgery and radiation therapy is doubtful. NSAIDs, most commonly indomethacin and sulindac, have been used in neo-adjuvant settings with partial responses or even complete resolution of desmoid tumours.^{17,18} Since extraabdominal desmoid tumours are usually positive for estrogen and progesterone receptors; thus, a role of hormonal therapy has been anticipated.¹⁹ Tamoxifen and toremifene have demonstrated a response rate of 30-50% over one year.²⁰ Mammary desmoids especially those associated with polyposis syndromes are characteristically negative for hormone receptors as was observed in our case and hence our patient was not offered any hormonal therapy.¹⁹ However, a study by Ishizuka et al. has shown tamoxifen to be effective even in Estrogen Receptor (ER) negative mammary desmoid tumors, partly explained by induction of synthesis of transforming growth factor beta-1 by ER-negative fibroblasts, which can cause apoptosis and regression of the tumor.¹⁹

A routine quarterly follow up is suggested for a minimum of three years as most of the local recurrences become apparent within this time frame.^{15,17} The patient in our case has been recurrence free after adjuvant radiotherapy despite posterior margin being positive after 1 year of follow up.

CONCLUSIONS

A high index of suspicion and a thorough work up is necessary to detect rare lesions like a desmoid tumor which can masquerade as breast carcinoma. Adjuvant radiotherapy is a viable treatment option to reduce local recurrence in cases when re-excision is not possible.

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Ethical approval: Not required

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