Fibromatosis of Breast: A Rare Entity and Review of Literature

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Abstract

Mammary fibromatosis is a rare and locally aggressive benign tumour of the breast. It originates from fibroblasts and myofibroblasts within the breast parenchyma and does not metastasize\textsuperscript{1}. The condition is locally aggressive and has high rate of recurrence. The etiology of fibromatosis is unknown. Breast imaging examinations often imitate breast cancer. Here we report a case of desmoids fibromatosis breast which mimicked carcinoma breast\textsuperscript{2}.

Keywords: Fibromatosis, Mesenchymal cells

Introduction

Breast fibromatosis is a very rare mesenchymal neoplasm (cell growth) that may occur in women, typically between ages of 25-45 years. Breast fibromatosis does not metastasize but may be locally aggressive and is prone to recur. So while it is not invasive breast carcinoma, it is still a serious tumour. This accounts for 0.2\% of all breast tumours they can be very painful. And if left untreated they can grow large and create unwanted cosmetic changes\textsuperscript{3}.

Mesenchymal cells are genetically immature cells. So this type of spindle like cell lesions such as fibromatosis, myofibroblastoma and nodular fascitis tend to form either from fibroblasts or myofibriblasts. Specialists also call this family of benign breast lesions stromal lesions, because they have formed out of supportive tissues and not glands or epithelial cells. They do not have an inflammatory component. If the lesion is clinically palpable it often presents as a movable hard or firm mass or lump. Skin retraction and fixation to muscle are frequently also present\textsuperscript{5}. Microscopically other lesions will tend to show an unencapsulated infiltrative growth of evenly distributed spindle cells mixed with collagenous material.

In the first place the cellular and mammographic appearance along with the rate of growth can readily be mistaken as spindle cell carcinoma. The exact cause of fibromatosis is not known. Hormonal imbalances are often suggested as contributing factor in various breast mass developments. Breast fibromatosis tends not to test positive for HER2 receptivity. Younger women of child bearing age tend to have fibromatosis...
which are more cellular, more mitotically active and with larger amount of mild cellular atypia. Some researchers feel that this condition arises denovo from within the breast parenchyma (functional stromal elements). However other researchers feel it instead arises denovo from the aponeurosis which overlay pectoralis muscle and moves into the breast parenchyma 6. Here we present a case of 65 years old female who presented with fibromatosis breast that mimicked carcinoma breast.

**Case Report**

A 65 year old woman presented with a palpable lump in the left breast. She initially noticed the lump 1 year back which was small in size and slowly increased to attain the present size of 8*5 cm. The lump was firm and was occupying all the quadrants and was felt below the nipple areolar complex. It was not attached to the nipple. There was no skin or muscle involvement. There was no associated nipple discharge. The axillary examination showed single discrete mobile node in the central group measuring 2*2 cm. No other nodes were palpable. She is a known case of T2DM on oral hypoglycaemic drugs for past 20 years. She was also on anti hypertensives. None of her first degree relatives had history of carcinoma breast. All routine blood investigations were normal except for marginally raised RBS. FNAC was done from the breast lesion and showed features suggestive of atypical ductal cell hyperplasia or ductal carcinoma in situ.

A modified radical mastectomy was done. The surgically resected specimen contained a mastectomy specimen which measured 16*15*6 cm. A grey white hard area was identified measuring 6.5*6.5 cm. (figure 1). The grey white hard area showed multiple whorled lobulated areas. Also a lymph node was recovered. Histopathological examination showed unencapsulated tumour tissue arranged in the form of long intersecting fascicles and shows infiltration into adjacent fat (Figure 2). Cells were bland looking spindle shaped with eosinophilic cytoplasm and entrapment of ducts with lining cells showing apocrine change. No mitosis, necrosis or atypia seen. Impression was benign spindle cell lesion or desmoids type fibromatosis 7. The patient was followed up for 6 months and the patient is disease free at the time of this case report.

![Figure 1: specimen of excised breast tissue](image-url)
Discussion

The world health organisation defines desmoids type fibromatosis as an intermediate soft tissue tumour that is characterised by clonal fibroblastic proliferation arising in the deep soft tissues with a tendency for infiltration of the local tissues and local recurrence but inability to metastasize. Cases of desmoids type fibromatosis are often broadly classified into two groups. The largest group contains sporadic group with incidence of 3.42 per million years forming 84-92 % of all cases. In this group somatic beta catenin activating mutations are considered to be cause of disease. The second smaller group of cases consists of those associated with familial adenomatosis polyposis. The etiology of mammary fibromatosis is unknown. It has no predilection for age, family history or exposure factors, although certain cases occur after trauma. The disease is usually painless and presenting symptom is always a lump. Breast imaging are not specific and mimic carcinoma. CT or MRI assists in defining the infiltration into adjacent tissue particularly in patients in whom there was a pre operative suspicion of chest wall involvement.

Standard treatment of this recurrent tumour involves wide surgical resection with safe margins. The reasons for such an aggressive surgical approach include the potential of fibromatosis to undergo aggressive local growth with high local recurrence rate when incompletely excised with positive margins. Mastectomy should be avoided in cases where it is possible especially in young women. Notable 24 cases of breast desmoids tumours that developed following augmentation mammoplasty have been described in literature.

A re assessment of the overall management of fibromatosis has taken place over the last few years and preservation of function has become a priority. The role of adjuvant radiotherapy also remains unclear. Patients should be followed carefully for atleast 3 years. MRI are best suited to accurately reflect disease progression and regression. In conclusion desmoids like fibromatosis is a rare breast neoplasm. Despite its classification as an intermediate soft tissue tumour breast fibromatosis possess the potential for aggressive local behaviour. Surgery remains a valid option. The present study reports a case with its clinical features and histological findings in order to improve and add to our knowledge of the disease.

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