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Rarely Diagnosed Cases of Fibrosarcoma Breast-A Case Series

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Abstract: *Breast sarcomas are rare tumors with an incidence of <1% of all breast malignancy [1]. Primary breast sarcomais defined as malignancy originating from mesenchymal tissue of the breast and does not include those arising from skin, muscle, and adjacent bones. They are similar to soft tissue sarcomas elsewhere in the body. Primary fibrosarcoma of the breast is a very rare tumor described in literature, accounting for 16% of all soft tissue sarcomas of the breast (2).Phyllodes tumours of the breast are commonly classified as benign tumours and rarely as borderline or malignant. Cases have been reported where originally benign tumours developed malignant features with recurrences [3]. Primary sarcomas of the breast are extremelyrare and account for less than 0.1% of all malignant tumours of the breast [4], of which the conversion of a phyllodes tumour to sarcoma of the breast is even rarer and only one such case has ever been reported [5].*

I. Introduction

Breast sarcomas are relatively rare accounting for 0.5 to 3% of malignancies in breast(9-15). These include fibrosarcoma, liposarcoma, alveolar soft part sarcoma, clear cell sarcoma, leiomyosarcoma, rhabdomyosarcoma, angiosarcoma, malignant fibrous histiosarcoma. Although, the most common breast sarcoma is cystosarcoma phyllodes, but it is not a true sarcoma (fibroepithelial lesion by definition) and is excluded from most recent studies on Primary Breast Sarcomas. Primary fibrosarcoma of the breast is a very rare tumor described in literature, accounting for 16% of all soft tissue sarcomas of the breast (2). In a review of the literature, Barnes and Pietruszka [6] reviewed all the cases of breast sarcoma that were seen at the Health Center Hospitals of the University of Pittsburgh during the 1945-1976 period, and they found that only 10 cases of primary breast sarcoma had been reported. Further, Adem et al. [7] reported that primary breast sarcoma comprised only 0.0006% of all breast malignancies presenting at his institution from 1940 to 1999.

However,only few cases have been reported till date regarding conversion of phyllodes into fibrosarcoma.Cystosarcoma phyllodes is a rare, predominantly benign tumor that occurs almost exclusively in the female breast.[8] Its name is derived from the Greek words *sarcoma* ("fleshy tumor"), and *phyllon* ("leaf"). Grossly, the tumor displays characteristics of a large, malignant sarcoma, takes on a leaflike appearance when sectioned, and displays epithelial, cystlike spaces when viewed histologically. Although the benign tumors do not metastasize, they have a tendency to grow aggressively and can recur locally.[9] Like other sarcomas, the malignant tumors metastasize hematogenously.

In 1981 [17] theWorldHealthOrganization adopted the term phyllodes tumor and as described by Rosen [18] sub classified them histologically as benign, borderline or malignant according to the features such as tumor margins,stromal overgrowth, tumor necrosis, cellular atypia, andnumber of mitosis per high power field The difficulty in distinguishing between fibroadenoma, benign phyllodes tumors, and malignant cystosarcoma phyllodes may be vexing for even the most experienced pathologist.[10] Reports suggest, however, that about 85-90% of phyllodes tumors are benign and that approximately 10-15% are malignant.[11]

Phyllodes tumors are rare fibroepithelial lesions. They makeup 0.3 to 0.5% of female breast tumors [14] and have an incidence of about 2.1 permillion, the peak ofwhich occurs in Women aged 45 to 49 years [15, 16]. The tumor is rarely found in adolescents and the elderly.

II. The Aim

this study is to report the rare cases of primary fibrosarcoma and the same arising out of malignant phyllodes tumor. Prognosis of these rare tumours is poor and treatment modality is not clearly defined. The rarity of this tumour and its successful treatment prompts us to report this case

III. Distinguishing Features

The characteristics of a malignant phyllodes tumor include the following [12]

- Recurrent malignant tumors seem to be more aggressive than the original tumor
- The lungs are the most common metastatic site, followed by the skeleton, heart, and liver
- Symptoms of metastatic involvement can arise from as early as a few months to as late as 12 years after the initial therapy
- Most patients with metastases die within 3 years of the initial treatment [13]

- No cures for systemic metastases exist
- Roughly 30% of patients with malignant phyllodes tumors die of the disease

IV. Etiology And Risk Factors

Phyllodes tumor is the most commonly occurring non-epithelial neoplasm of the breast, although it represents only about 1% of tumors in the breast.[9] It has a smooth, sharply demarcated texture and typically is freely movable. It is a relatively large tumor, with an average size of 5cm. However, lesions of more than 30cm have been reported. The etiology of phyllodes tumors is unknown. It has been suggested that, in a proportion of fibroadenomas, a somatic mutation can result in a monoclonal proliferation, histologically indistinguishable from the polyclonal element, but with a propensity to local recurrence and progression to a phyllodes tumor which has also been

supported by clonal analysis. It has also been postulated that stromal induction of phyllodes tumors can occur as a result of growth factors produced by the breast epithelium.

Trauma, lactation, pregnancy, and increased estrogen activity occasionally have been implicated as factors stimulating tumor growth. The nature of these factors is unclear but endothelin-1, a stimulator of breast fibroblast growth may be significant.

Pathogenesis

Unlike carcinoma breast, phyllodes tumors start outside of the ducts and lobules, in the breast's connective tissue, called the stroma which includes the fatty tissue and ligaments that surround the ducts, lobules, and blood and lymph vessels in the breast. In addition to stromal cells, phyllodes tumors can also contain cells from the ducts and lobules.

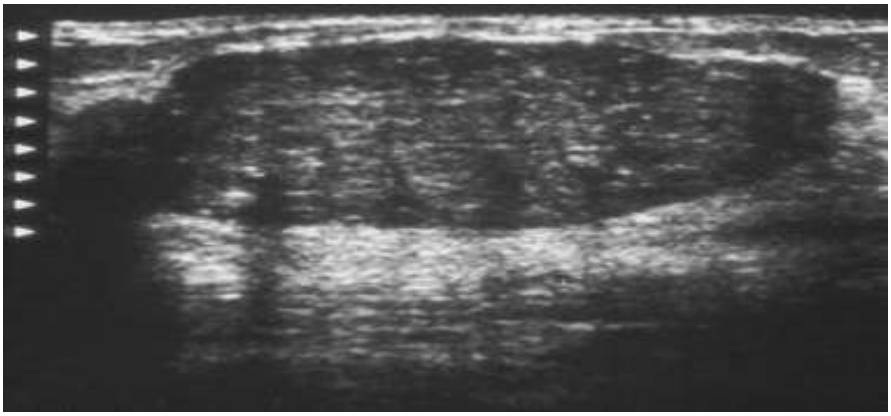
Clinical features

Case Series

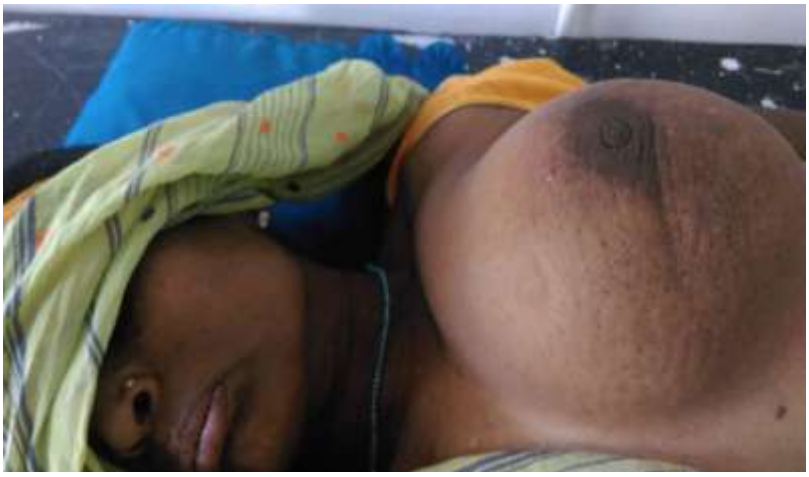
Case 1:

40 years old female presented with chief complaints of painless breast lump 10 months ago. The patient incidentally detected the mass 10 days prior to the clinical visit. She had no history of breast abnormalities, previous breast surgery or radiation therapy. Her mammography revealed large lobulated dense opaque mass as shown below

USG shows heterogenous hypoechoic, well marginated lesion



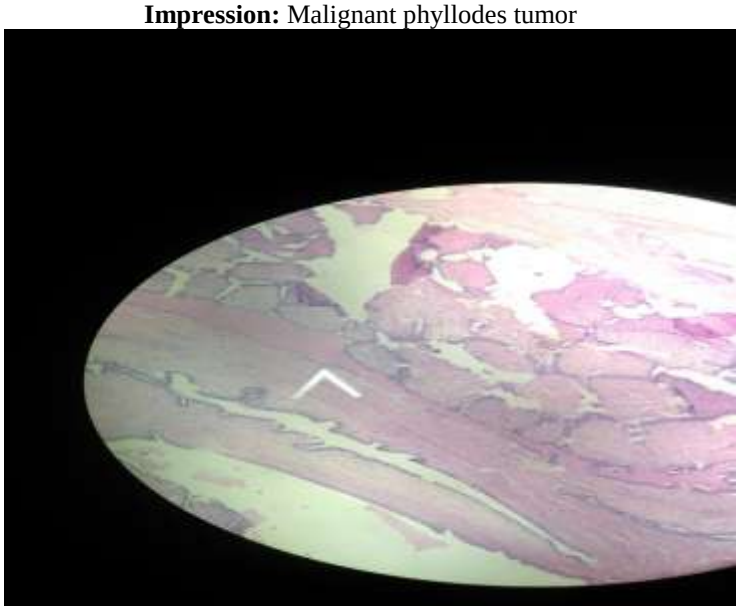
O/E- 8*7cm relatively well-defined mass in the upper outer quadrant of the left breast. There was no overlying skin abnormality or palpable axillary or supraclavicular lymph nodes.



Preliminary FNAC findings revealedhypercellular smear with dispersed pleomorphic spindle cells predominantly showing moderate nuclear atypia and prominent nucleoli..the smear showed predominantly stromal component with minimal ductular component.
IMPRESSION:- suggestive of malignant phyllodes tumor

After this diagnosis patient underwent lumpectomy and tissue was sent to our department for biopsy reporting. The histopathological examination revealed infiltrative margins, stromal hypercellularity and overgrowth of the glands by the sarcomatousstroma.the neoplastic stromal component showed marked nuclear atypia and numerous mitoses(>10hpf).

Impression: Malignant phyllodes tumor



The same female presented at our institution ten months after the lumpectomy. She complained of mild pain at the operative site. O/E- a healthy scar was present with mild induration. No palpable lymph nodes were present. No abnormality was detected in the right breast and the axilla .Patient was again advisedFNAC which revealed no ductal cells and only mesenchymal component with increased atypia and mitoses.

Patient was advised wide excision biopsy and histopathological examination of the given section revealed a highlycellular, spindle cell tumor that displayed an interdigitating fasciculated growth pattern, the so-called herringbone pattern .The tumor cells were predominantly spindle shaped cells that showed little anisopoikilocytosis along with scanty cytoplasm with indistinct cell borders. The nuclei showed mild to moderate atypia. The mitotic activity was up to 6-10 per 10 HPF.

Impression- Fibrosarcoma

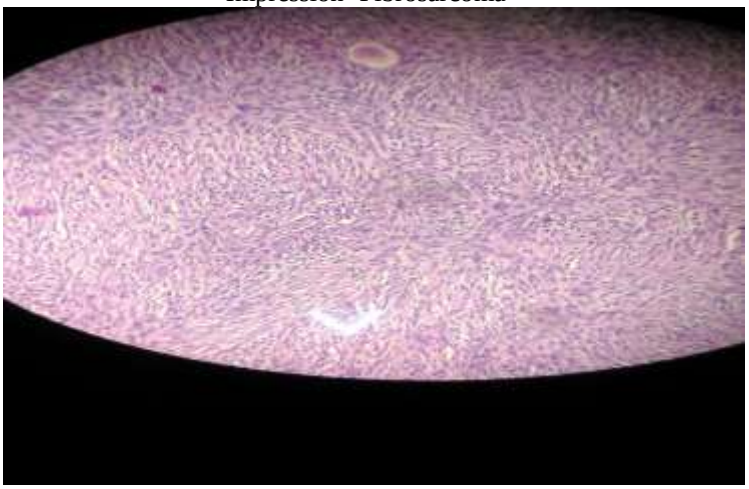
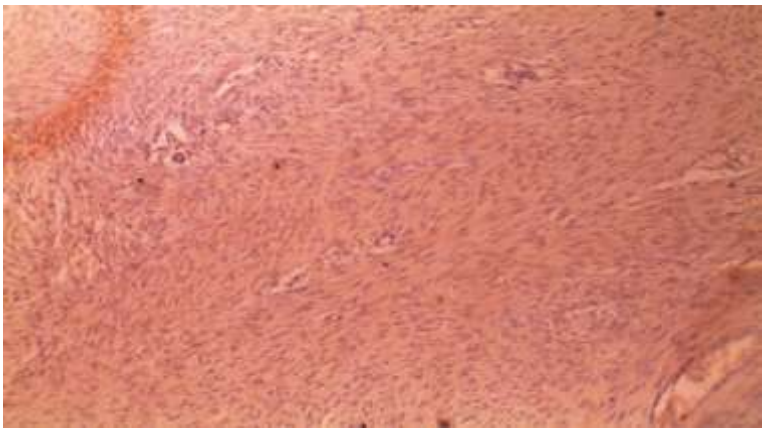


Fig-H&E stained section of Secondary Fibrosarcoma arising out of malignant phyllodes tumor.

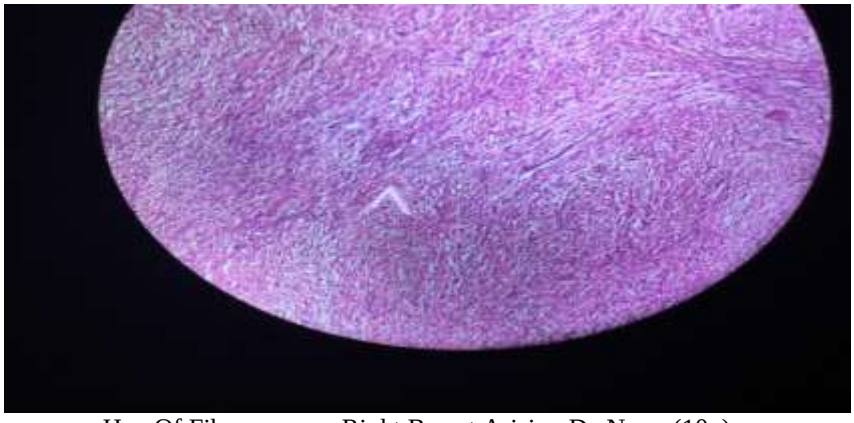
The diagnosis was further confirmed by Immunohistochemistry staining of the given section by vimentin which showed positivity confirming the diagnosis of fibrosarcoma.

Ihc Study Showing Vimentin Positivity In Fibrosarcoma

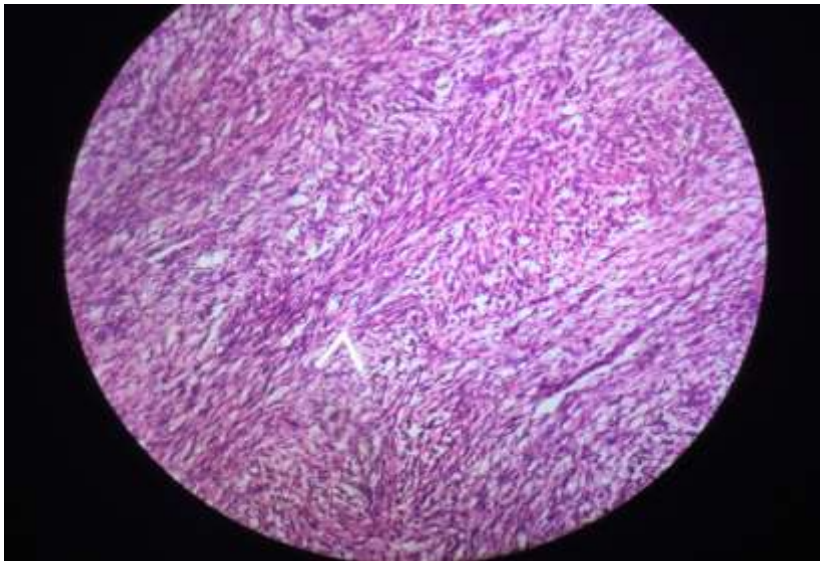


CASE 2:- A Case of fibrosarcoma of rightbreast arisingdenovo.35 years female presented with chief complaints of painless large lump right breast involving all quadrants since 2 months which gradually increased in size.no systemic complaints or significant past history.

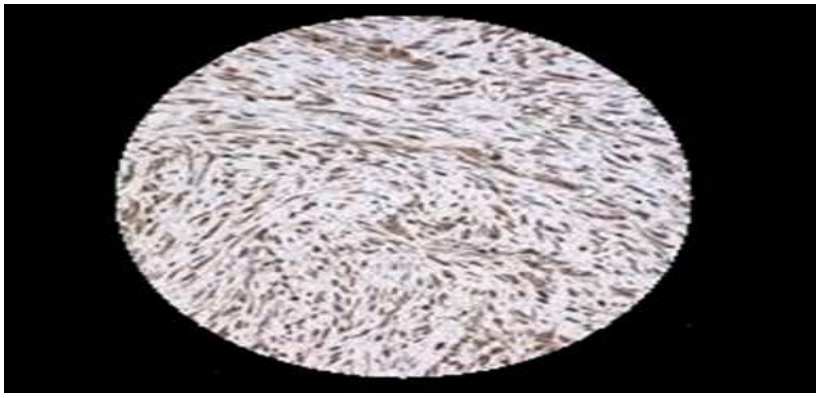
O/E:-non –tender lump measuring 10*8cms located in right breast involving all the four quadrants.no ulceration present over the skin overlying the lump. Preliminary FNAC findings revealed highly cellular smear with atypical spindle cells against the background of loose fragments of fibrous stroma and bare nuclei. Wide excision biopsy on first hand revealed FIBROSARCOMA which was subsequently confirmed by vimentin positivity.



Hpe Of Fibrosarcoma Right Breast Arising De Novo (10x)



Hpe Of Fibrosarcoma Right Breast Arising De Novo(40 X)



Diffrential Diagnosis

Liposarcoma,alveolar soft part sarcoma, clear cell sarcoma, leiomyosarcoma, rhabdomyosarcoma, angiosarcoma, malignant fibrous histiosarcoma, osteosarcoma,chondrosarcoma

Investigations

Complete Blood Count with Erythrocyte Sedimentation Rate,Serum uric acid,Renal Function Test,Liver Function Test,Chest x-ray,CT SCAN,MRI,2D Echocardiography ,Mammogram,FNAC.

V. Result

Fibrosarcoma is a rare breast lesion barely diagnosed .the first case was conversion of malignant phyllodes tumor to fibrosarcoma of left breast over a period of ten months .The second case is a recently diagnosed case of fibrosarcoma of right breast arising denovo.

VI. Discussion

The fibrosarcoma is a rarely diagnosed tumor in breast. The interval between the diagnosis of the primary tumour and identification of metastasis ranged from seven months to five years, whereas the interval between recurrent tumour and metastasis was from 6 to 24 months or in some cases metastasis were present simultaneously [3]. Very rarely, the malignant potential increases and results in the conversion to a sarcoma of the breast. Fibrosarcomas are amongst some of the most rare tumours of the breast. Any breast neoplasia that does not display characteristics of a fibroadenoma are designated stromal sarcoma of which fibrosarcomas are a small percentage.

Distant metastases are developed in3.2%, 11.1% and in 28.6% of patients with benign, borderline and malignant phyllodes tumours, respectively [9]. With histopathological

conversion to a sarcoma, the chances of distant metastases increase.

Diagnosis And Treatment:- Accurate preoperative pathological diagnosis allows correctsurgical planning and avoidance of reoperation, either to achieve wider excision or for subsequent tumor recurrence[19-21]. Treatment can be either wide local excision or mastectomy provided histologically clear specimen margins are ensured.(22,23)

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