

Stromal tumours of the breast

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ABSTRACT

Introduction: Phyllode tumours (PT) are fibroepithelial lesions that account for less than 1% of all breast neoplasms. With the non-operative management of fibroadenomas widely adopted, the importance of PT today lies in the need to differentiate them from other benign breast lesions. The study was done to study the spectrum of various stromal tumours of the breast and the age distribution of the cases affected.

Materials & Methods: A retrospective and prospective study was done on patients who were referred for lump in the breast. Cases were analyzed in detail regarding complete history, family history, clinical examination and other findings.

Results: Stromal tumours of the breast were studied for a period of two years. Out of the total 19 cases, benign PT were 11 cases, borderline PT were 05 and one case each of malignant PT, leiomyoma and neurofibroma (NF) were diagnosed.

Conclusion: The present study highlights the spectrum of various stromal tumors of the breast like leiomyoma, neurofibroma and malignant PT. Complete excision of malignant PT should be done to avoid distant metastasis and the importance of fine needle aspiration cytology (FNAC) in early diagnosis of the tumour.

Keywords: Stromal tumours, Malignant phyllode, Leiomyoma, Neurofibroma of breast

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INTRODUCTION

Phyllode tumours are fibroepithelial lesions that account for less than 1% of all breast neoplasms.¹ With the non-operative management of fibroadenomas widely adopted, the importance of PT today lies in the need to differentiate them from other benign breast lesions. Although PTs may have been described as early as 1774, the lesion was first fully characterized in 1838 by Johannes Müller.² The term cystosarcoma phylloides was chosen to emphasize the leaf like pattern and fleshy gross appearance of the lesion. Among the many other names subsequently applied to the tumour, the only ones currently used are periductal stromal tumour and phylloides tumour. The former term was proposed to emphasize the probable origin from the specialized periductal stroma, but phylloides tumour has become the preferred nomenclature. The

diagnosis of PT should always include subclassification as benign, borderline and malignant. The distinction among these three subgroups is based on the histologic characteristics of the tumours and is predictive of the probable clinical course. Stromal tumours of the breast are very important to diagnose because tumours like malignant PT may metastasize to various organs like lungs, brain, etc. Careful examination on FNAC and histopathology helps in reaching an early diagnosis and proper treatment.

MATERIALS AND METHODS

A retrospective and prospective study of stromal lesions of the breast were studied for a period of two years. FNAC was done and confirmed by histopathology. The breast lump examination was done carefully and also the involvement of any lymph node was assessed. Complete family history

and menstrual history was taken. FNAC was done under all aseptic conditions by 23 gauge needle attached to 10 ml disposable syringe. Slides were air dried, fixed, with ether alcohol and stained by Haematoxylin & Eosin and Giemsa stains.

RESULTS

Out of the total 19 cases, Benign PT were 11 cases, Borderline PT were 05 and one case each of malignant PT, leiomyoma and neurofibroma (NF) were diagnosed (table 1). The age group distribution of the cases was shown in table 2. Maximum numbers of cases (11) were between the age of 41 – 50 years, followed by 06 cases in the age of 31-40 years. Two cases were in the age group of 21-30 years.

Table 1: Spectrum of various stromal tumours

Lesion	Number of cases
Benign PT	11
Borderline PT	05
Malignant PT	01
Neurofibroma	01
Leiomyoma	01
Total	19

Table 2: Age distribution of the cases

Age group	Number of cases
21-30	02
31-40	06
41-50	11
Total	19

DISCUSSION

PT previously called cystosarcoma phyllodes are the fibro epithelial tumours of the breast representing 2-3% of all fibroepithelial tumours and less than 01% of all breast tumours.¹ At least 12.5% of patients with PT have a concurrent diagnosis of a benign fibroadenoma.³ The pathological appearance of stromal overgrowth, stromal cellularity, degree of nuclear atypia, number of mitotic cells and the nature of the tumour margin determines whether a tumour is a benign fibroadenoma, benign PT, borderline PT or malignant PT. The stroma from malignant PT is distinguished by marked cellularity with nuclear atypia, nuclear pleomorphism, increased mitotic activity and stromal cell overgrowth.^{4,5} Heterogenous stromal components are seen in PT and is commonly associated with malignant PT.

Phyllode tumour of the breast has biphasic histological features with both epithelial and stromal

components. Careful examination of the stromal component is essential since it is the pathological features of the stromal cells that determine its malignant potential. Clinically the patient presents with a firm to hard discrete palpable tumour. There are no specific clinical features that reliably distinguish among a fibroadenoma, benign PT and malignant PT.⁶

The diagnosis of PT may be favoured if the tumour is larger than 4 cm, or if there is a history of rapid growth. Origin in a pre existing fibroadenoma or malignant transformation of a benign PT is suggested when the patient reports enlargement of a pre existing tumour that was previously stable for a number of years. A fibroadenoma may sometimes evolve into PT as suggested by clonal analysis of three tumours. Initially, these tumours were diagnosed as fibroadenoma and recurred as PTs.⁷ The tumours were studied for evidence of trinucleotide repeat polymorphism of the X chromosome linked androgen receptor (AR) gene and random inactivation of the gene by methylation. It was observed that the same allele of the AR gene was inactivated in the fibroadenoma and PT samples from each patient. This result was very unlikely to occur in three separate cases and can be attributed by chance.

Here we present a case of malignant PT. A 59 year old female presented with a large left breast mass of size 12 x 10 cm, firm to hard since 3 years. The swelling was gradually increasing and painful from past one year. FNAC was done and we reported it as malignant PT comprising of highly cellular smears showing marked pleomorphic and hyperchromatic nuclei with prominent nucleoli, necrosis and hemorrhage [Figure 1]. Total mastectomy was done for this patient and on gross we noticed cartilage and bony tissues along with focal areas of necrosis [Figure 2] and it was confirmed on histopathology as malignant PT [Figure 3 & 4].

We also present a case of neurofibroma of the breast, which is a very rare presentation among breast tumours. NF's are benign nerve sheath tumors, which were first described by Smith in 1849 and later by Von Recklinghausen in 1882. The majority of NF are solitary lesions that occur in the dermis or subcutis and are evenly distributed over the body surface.^{8,9} Solitary NF are rare and here we present a case of 38 year female with lump in right breast of size 5X3 cm since 6 months. FNAC was done and reported as spindle cell lesion [Figure 5a]. Excisional biopsy [Figure 5b] was performed for the same and microscopy showed tumour tissue comprising

mainly of elongated and spindle cells having wavy and tapering nuclei and confirmed as neurofibroma [Figure 6].

Leiomyoma is a benign smooth muscle neoplasm. They can occur in any organ. Leiomyoma of the breast is a very rare non epithelial tumour.¹⁰ Here we present a case of 50 year female who presented to us with a painless right sided breast lump of size 5X4

cm since one year in right upper quadrant, firm and mobile. FNAC was performed and show spindle cells arranged in whorling pattern and in sheets and reported as spindle cell lesion [Figure 7a]. Excisional biopsy was done [Figure 7b] and histologically showed bundles of smooth muscle tissue arranged in whorls and in fascicles and it was confirmed as leiomyoma of the breast [Figure 8].

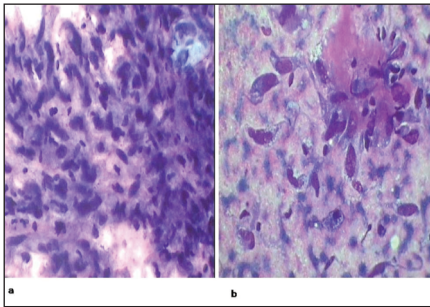


Figure -1

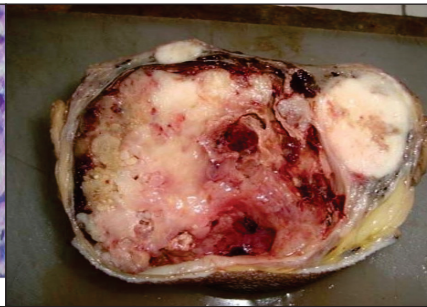


Figure -2

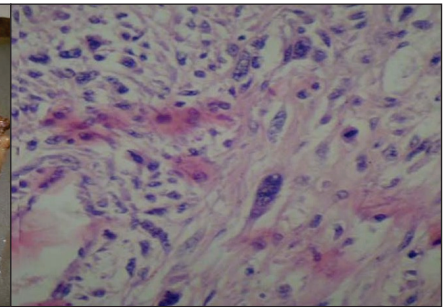


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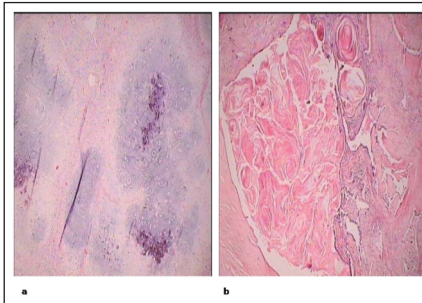


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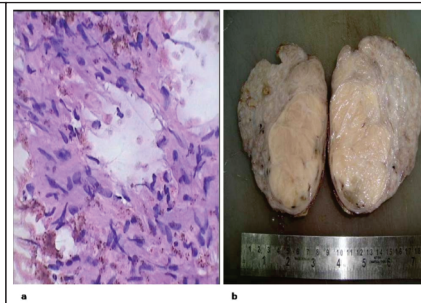


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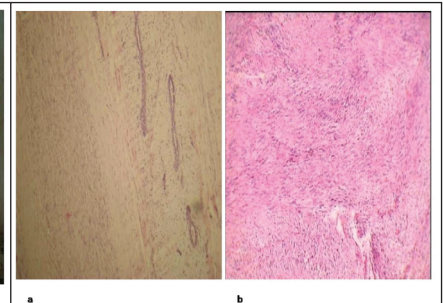


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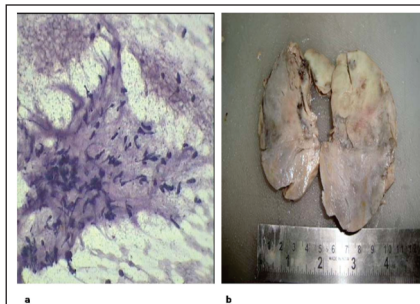


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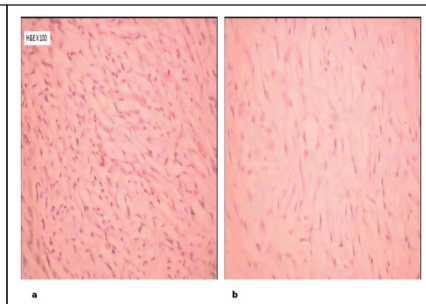


Figure -8

Figure legends:

Figure 1: FNAC smears showing cells with marked pleomorphic and hyperchromatic nuclei with inconspicuous nucleoli [Figure a, H&E, x40]. Areas of necrosis also seen [Figure b, H&E, x40].

Figure 2: Cut section of breast showing focal areas of cartilage, bone, necrosis, hemorrhage along with tumour tissue.

Figure 3: Section showing marked pleomorphic and hyperchromatic nuclei with prominent nucleoli and occasional mitotic figures [H&E, x40].

Figure 4: Section showing tumour tissue along with cartilaginous areas [Figure a, x 40 H&E]. Section showing necrotic and hemorrhagic areas [Figure b, H&E, x40].

Figure 5: FNAC smears showing spindle shaped cells with elongated and tapering nuclei [Figure a, H&E, x40]. Gross showing tumour of size 4x3cm, grayish yellow. [Figure b].

Figure 6: Section showing tubules, ducts and tumour tissue comprised of proliferation of smooth muscle tissue arranged in whorls and fascicles [Figure 5a & b, H&E, x40]

Figure 7: FNAC smears showing proliferation of spindle shaped cells with wavy nuclei [Figure a, H&E, x40]. Gross showing breast of size 5x3cm, cut section gray white to yellow [Figure b].

Figure 8: Section showing tumour tissue comprising of spindle shaped cells having wavy and tapering nuclei in a myxoid background [Figure a & b, H&E, x40].

The fundamental principle of therapy of PTs is complete excision to prevent local recurrence.¹¹. Features that predisposes to local recurrence are incomplete excision, an invasive tumour border, and secondary tumour nodules at the periphery. Primary tumour size may be a factor in the success of local excision, because a more generous margin may be possible when tumours are small. Because the diagnosis of PT is not anticipated clinically in many cases, surgical excision may be initially incomplete and re-excision is required. The primary excision and re-excision specimens should be inked and margins thoroughly examined histologically. Mastectomy is indicated as primary therapy if a malignant PT cannot be encompassed with a cosmetically acceptable incision. Axillary lymph node mapping or dissection is appropriate if there is concurrent carcinoma in the PT or elsewhere in the same breast or if the lymph nodes appear to be clinically involved by the tumour.

The overall 5 year survival rate for PT is about 90%.¹² Local recurrences, which occur in about 30% of cases, and metastases that develop in about 10% of cases are usually detected within 3 years of primary treatment, although occasional instances of late recurrence have been reported. Most deaths due to metastatic PT occur within 5 years of diagnosis.

The main aim of our study was to highlight the role of FNAC and stromal tumours of breast like neurofibroma, leiomyoma and malignant PT which needs complete excision to prevent metastasis and thereby mortality among the cases.

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