Research Paper

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Malignant Phyllodes Tumor: A Rare Presentation

Medical Science

KEYWORDS : phyllodes, tumor, malignant

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ABSTRACT

Introduction. Phyllodes tumours are rare fibroepithelial lesions.Accurate preoperative pathological diagnosis allows correct surgical planning and avoidance of reoperation. Treatment can be either wide local excision or mastectomy to achieve histologically clear margins. Discussion. The exact aetiology of phyllodes tumour and its relationship with fibroadenoma are unclear. Women aged between 35 and 55 years are commonly involved. The median tumour size is 4 cm but can grow even larger having dilated veins and a blue discoloration over skin. Palpable axillary lymphadenopathy can be identified in up to 10–15% of patients but <1% had pathological positive nodes.Mammography and ultrasonography are main imaging modalities. Cytologically the presence of both epithelial and stromal elements supports the diagnosis.The value of FNAC in diagnosis of phyllodes tumour remains controversial. Surgical management is the mainstay and local recurrence in phyllodes tumours has been associated with inadequate local excision. The role of adjuvant radiotherapy and chemotherapy remains uncertain and use of hormonal therapy has not been fully investigated. Conclusion.The preoperative diagnosis and proper management are crucial in phyllodes tumours because of their tendency to recur and malignant potential in some of these tumours.

INTRODUCTION

The tumor initially named cystosarcoma phyllodes is now generally known as phyllodes tumor. It is rare fibroepithelial neoplasm characterised by proliferation of stromal and epithelial cells and account for less than 1% of all breast tumor. It occur occur mostly between age group of 40-50 years. Larger the phyllodes tumor, more is the chances of its being malignant but assessment of its biological behaviour is based on histological features includes stromal cellularity, atypia, mitosis, infilterative growth, stromal overgrowth, presence or absence of necrosis. Phyllodes tumor exhibit an enhanced intracanalicular growth pattern that is leaf like projection into dilated lumen. Malignant stromal transformation in Phyllodes tumor is usually fibrosarcomatous type but sometimes the mesenchymal component may show areas resembling sarcoma, chondrosarcoma, liposarcoma, leiomyosarcoma, rhabdomyosarcoma.

CASE REPORT

51 year female presented in emergency department with complaints of pain in right breast since 15 days and increasingly palpable mass in the right breast since 2 years. General examination revealed no abnormality.

Local examination of the right breast showed enlarged laterally displaced hard mass in the right upper quadrant with engorged dilated veins and retraction of nipples. No lymph node was palpable. Haematological and biochemical results were within normal limits. A clinical differential diagnosis of cystosarcoma phyllodes and fibroadenoma was given. Ultrasonography and chest x ray were inconclusive. The left breast and axilla was normal. The respiratory and abdominal examination revealed no abnormality. High resolution computated tomography showed multiple well defined large cyst in the anterior segment of right upper lobe. Fine needle aspiration cytology showed malignant phyllodes. Right mastectomy was done and was received in histopathology in 10% formalin solution. The received specimen of right breast weighing 1.3 kg and measuring 18x6x12cm with skin flap measuring 13x11x4cm and tumor proper was measuring 11x10x6cm. Cut section shows variegated appearance showing with cystic areas along with nodules showing areas of necrosis. Final histopathology report of malignant phyllodes tumor was given.

Figure 1: Showing lobulated, whitish yellowish showing cytic as well as firm areas.

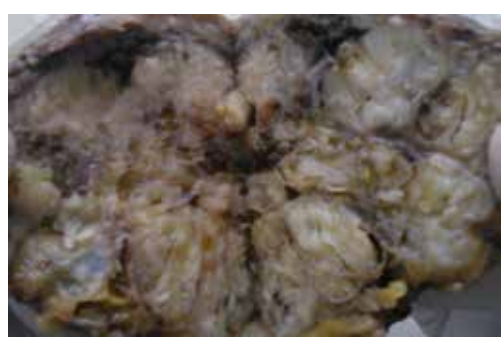

Figure 2: Showing lobulated yellowish white areas along with areas of hemorrhage

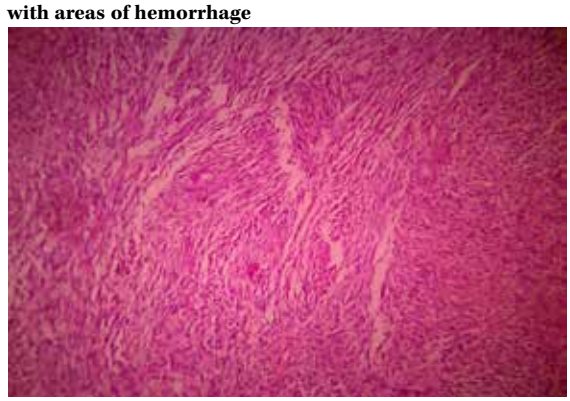

Figure3:

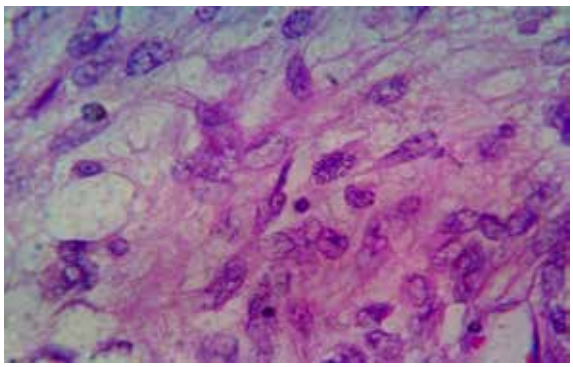

Figure 4:

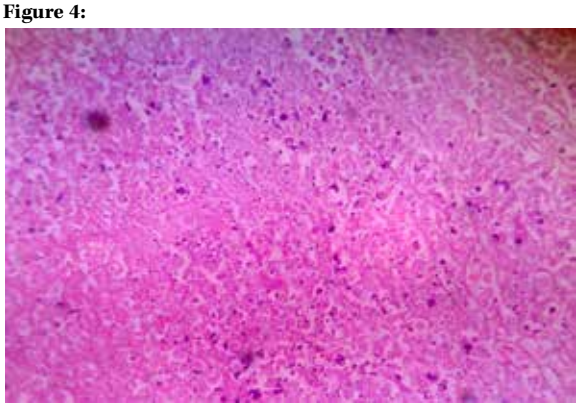

Figure 5:

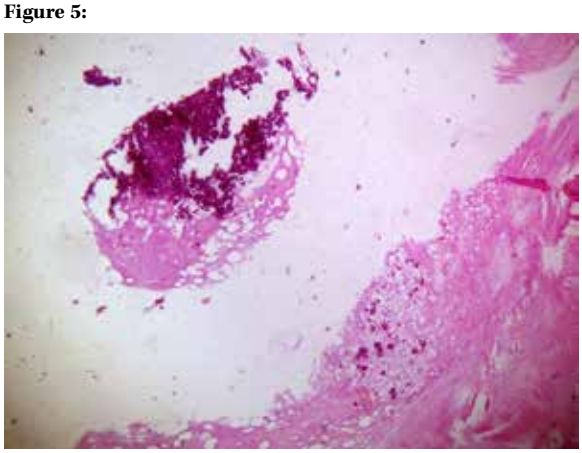

Figure 6: DISCUSSION

A phyllodes tumor is a rare distinctive fibroepithelial tumor of the breast. It constitutes 1 % of all the breast tumors1. The tumor was first described by Muller in 19382. Muller described cystosarcoma phyllodes as leaflike masses with a cystlike shape and complete envelopes. Recurrent malignant tumor seems to be more aggressive than the original tumor. The lungs are the most common metastatic site, followed by the skeleton, heart, and liver. Most patients with metastases die within 3 years of the initial treatment3. Depending on the cellularity, invasiveness of the tumor, cytological atypia, and mitotic count, phyllodes tumor is classified into benign and malignant. Malignant is further divided into low-grade and high-grade based on histology. a malignant phyllodes tumor shows marked stromal hypercellularity, marked cellular pleomorphism, numerous mitotic figures (>10 /10 high power fields), an infiltrative margin and stromal overgrowth. A phyllodes tumor which displays indeterminate features is categorized as a borderline malignancy.The disease occur predominatly in middle age women, with average of 40years. They often present clinically painless mass with average size of 4-5 cm. Axilliary lymph node metastasis is rare. Local recurrence for benign is 21%, borderline 46% and malignant 65%.pts, also known as cystosarcoma phyllodes, were first named by Johannes Muller4,5. At present, the exact causes of PTs remain unknown, although the majority of researchers consider that PTs are likely to have similar epidemic factors to fibroadenoma of the breast, which is associated with estrogen secretion and metabolic disorders. In 1982, PTs were classified by the WHO as benign, borderline and malignant, on the basis of cell density, atypia, mitotic figures, tumor borders and hemorrhagic necrosis. Malignant tumors (either low or highgrade) are common in elderly females and exhibit frequent metastasis (lymphatic and bloodborne) with a high mortality rate. Due to the overlapping histological and clinical features between small PTs and fibroadenomas, there are no known tumor markers or blood tests to aid in the confirmation of the diagnosis. Consequently, it is easy to misdiagnose the majority of PTs as fibroadenomas. The differentiation of PTs from benign fibroadenomas is difficult using ultrasound, mammograms and magnetic resonance imaging (MRI). The study suggested that the size, cellularity of stromal fragments and proportion of spindle cells in the background are significant features in such differentiation. Spindle cells appear to be present in large numbers in PTs compared with fibroadenomas.

Paddington Clinicopathological Suspicion Score. This outlines criteria to assist in the selection of patients for core biopsy, for use in conjunction with existing local protocols. The aim of developing the score is to improve the rates of preoperative diagnosis.

Clinical findings:-

(i) Sudden increase in size in a longstanding breast lesion
(ii) Apparent fibroadenoma >3cm diameter or in patient >35 years

Imaging findings

(i) Rounded borders/lobulated appearance at mammography
(ii) Attenuation or cystic areas within a solid mass on Ultrasonography

FNAC findings:-

(i) Presence of hypercellular stromal fragments
(ii) Indeterminate features

ANY 2 features mandate core biopsy.

Classification

WHO Criteria

World Health Organization divided phyllodes tumor into benign, borderline, and malignant categories based on the degree of stromal cellular atypia, mitotic activity per 10 high power fields, degree of stromal overgrowth (these three are main), tumor necrosis, and margin appearance.

Criteria	Benign	Borderline	Malignant
Stromal cellularity and atypia	Minimal	Moderate	Marked
Stromal overgrowth	Minimal	Moderate	Marked
Mitoses/10 high power fields	0–4	5–9	≥10
Tumor Margins	Well circumscribed with pushing tumor margins	zone of microscopic invasion around tumor margins	infiltrative tumor margin

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Criteria Proposed by Azzopardi et al.6 and Salvadori et al.[7]

Criteria	Histological type		
	Benign	Borderline	Malignant
Tumor margins	Pushing	--	Infiltrative
Stroma cellularity	Low	Moderate	High
Mitotic rate (per 10hpf)	<5	5–9	≥10
Pleomorphism	Mild	Moderate	Severe

Role of Tumor Markers in Phyllodes Tumor. Increased p53 protein and Ki-67 antig en expression has been detected in malignant phyllodes tumors and they may be valuable in differentiating fibroadenomas from phyllodes tumors. Furthermore, in phyllodes tumors, p53 and Ki-67 expression has been shown to correlate with negative prognostic factors.

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