www.jmscr.igmpublication.org
Impact Factor 5.84
Index Copernicus Value: 71.58
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v5i9.105

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Malignant Phyllodes Tumour of Breast: Case Report and Review of Literature

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ABSTRACT
Phyllodes tumors are rare fibroepithelial breast tumors, which are sometimes difficult to preoperative diagnoses and have unpredictable clinical outcome. Malignant transformation of phyllodes is extremely rare. These tumors must be suspected in patients with rapid-growing breast nodules, with a history of local recurrences to avoid inappropriate management.
We report a case of a large phyllodes tumour of breast with malignant transformation in a 45 year old lady.

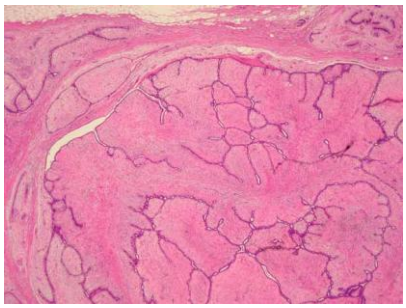
Introduction

The phyllodes tumor was originally described by Johannes Muller in 1838. Classically, the name cystosarcoma phyllodes was assigned because of the tumor's fleshy appearance and tendency to contain macroscopic cysts. The term, however, this term is a misnomer as these tumors are usually benign. Phyllodes tumor is the currently accepted nomenclature according to the World Health Organization (WHO) [1].

Phyllodes tumors are biphasic fibroepithelial neoplasms of breast[1] which are generally classified as benign, borderline and malignant[2]. Borderline tumors have great potential for local recurrence. They make up 0.3 to 0.5% of female breast tumors[4] and have an incidence of about 2.1 per million.approximately 85–90% of phyllodestumours are benign and 10–15% were malignant[12]

Phyllodes tumors are sharply demarcated from surrounding tissue which is compressed.

Connective tissue composes of the bulk of these tumors which have mixed gelatinous, solid and cystic areas. Most malignant phyllodes tumors contain liposarcomatous/rhabdomyosarcomatous elements rather than fibrosarcomatous component.



Evaluation of number of mitosis may help in diagnosis of malignant tumor [2]. Few reports have specifically commented on the giant phyllodes tumor, an entity that presents the surgeon with several unique management problems. The cut off point for designation as a giant phyllodes tumor is 10 cm[3].

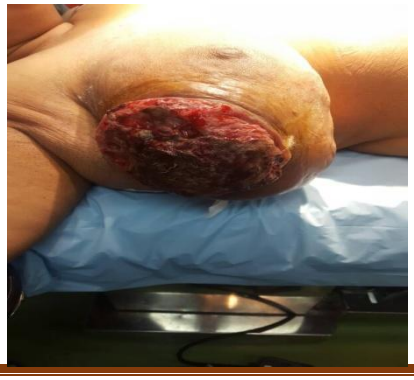
Usually wide local excision of the tumor with 1 cm[2] breast margin may suffice as the treatment but larger malignant variety may require tailoring of treatment pertaining to that individual case and number of recurrences. Radiotherapy may have a role in the treatment of phyllodes tumor depending of the number of recurrences, mitotic index, bulky tumor, status of the resection margins p53 and Ki67 expression[5].

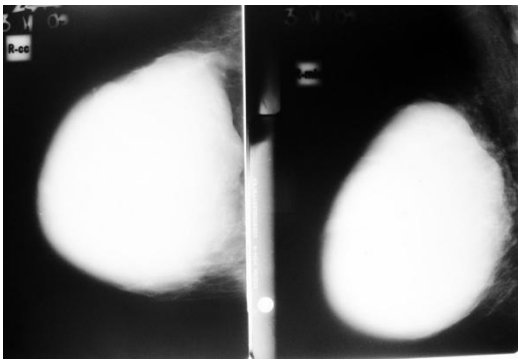
Case Report

A 45 year-old, perimenopausal female, presented with a large fungating mass in the right breast. It was associated with severe pain and fever since 2 days. At first the patient had a small lump in the right breast which enlarged over the course of one year and then became fungating. There was no personal or family history of breast cancer. Her menstrual history and obstetric history were unremarkable.

Physical examination findings were of a protruding and hardened palpable mass occupying the upper and lower outer quadrants of the right breast, There was an overlying ulcer of 15x10 cm with everted edges and lobulated surface with areas of active bleeding and slough, suggestive of cutaneous involvement. There was clinically no axillary lymphadenopathy.

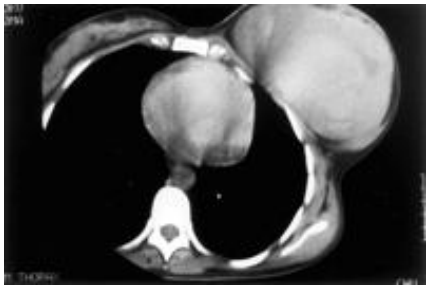
Mammogram showed multiple, hyperdense rounded masses with well-defined margins in the right breast with axillary lymphadenopathy. Ultrasound evidenced in the same topography; rounded confluents masses of well-defined and regular contours, with heterogeneous echo texture and cystic areas in its interior, posterior acoustic enhancement and internal flow on color-flow Doppler examination. (BIRADS: IVc).





Ultrasound-guided percutaneous biopsy (core biopsy) was performed on the breast lesion, the histopathologic examination showed biphasic neoplasm with malignant stromal component.

Thorax x-rays and abdominal ultrasound were performed for staging, which showed no significant alterations. Because of the malignant result on core biopsy, a PET-CT was ordered to rule out systemic spread of the disease. Also, CT images helped in better delineating the local extent of the disease. PET-CT revealed a heterogenously enhancing FDG avid lesion (SUV Max: 3.36) of 12.3 x 15.2x12.7 cm with perilesional fat stranding. The fat planes with pectoralis muscle were focally lost but there was no obvious chest wall infiltration. Perilesional low grade FDG avid satellite nodules (SUV Max: 1.58) were noted. Another low grade lesion of 3.8x3.6x2.5 cm was noted inferior to the above lesion, retroareolar in position. The skin infiltration was appreciated as low grade cutaneous thickening of 1.3 cm (SUV Max: 1.98). There were low grade metabolically active sub-centimeter lymph nodes in right axilla.



The patient was counseled regarding the malignant nature of disease and she underwent a modified radical mastectomy with axillary dissection. Intra-operatively a part of pectoralis muscle was shaved off to achieve adequate

resection margins. The histopathology report was of malignant phyllodes tumor with reactive lymph nodes (0/11). The specimen measured 23x16x15 cm with fungating fleshy tumor of 17x17x14 cm located in all quadrants. The cut resection margin was 2.8 cm from the base.

The further line of management after a multidisciplinary tumor board meet was to give radiotherapy in this patient.

Discussion

Phyllodes tumors are rare lesions with an incidence of less than 1% of all breast tumors[1]. Its incidence is greater in white women 35-55 years of age[1]. Clinically, phyllodes tumors are commonly present as a nodule, mobile, painless and with rapid growth. Histologically, these tumors are biphasic lesions consisting of a stromal and epithelial components, arranged in an undulating configuration with many slit-like spaces and crevices surrounded by an increased growth of mesenchymal cells[6]. The 2003 WHO tumor classification has proposed that phyllodes tumor be classified into three categories benign, borderline and malignant[7] according to the degree of cellular atypia, mitotic activity, characteristics of the tumor margins and the presence of stromal growth[8]

There are no mammographic abnormalities or ultrasonographic pathognomonic signs. In mammography, these lesions commonly present as voluminous isodense mass to breast parenchyma, circumscribed, which may be associated with calcifications. In ultrasound, they are generally characterized as a solid lobulated nodule of well-defined contours, and may be associated with cystic components.[9]

Phyllodes tumors present rapid growth, however, when in smaller dimensions, it's difficult to differentiate them from fibroadenoma, including histopathologic aspects of fragments obtained through core biopsy, where the main criterion for differentiation of fibroadenoma is the higher stromal cellularity presented in phyllodes tumors. Problems occur when characterizing malignant forms due to their large cellularity and atypia which varies, making broader samples necessary for conclusive diagnosis, even in surgical specimens. Mitotic count may also be negatively affected by the size of the fragments obtained by core biopsy and in the malignant shapes, the stromal overgrowth can result in the absence of epithelial part in the sample.[10] The treatment for phyllodes tumors remains surgical removal of this tumor. It is essential to keep a sufficient margin of healthy tissues, which reduces the risk of local recurrence. For borderline or malignant phyllodes tumors or in local tumor recurrence, mastectomy may be considered. The role of adjuvant treatments is unproven and must be considered on a case-by-case basis. It is necessary to follow up the patients, because there is a risk of local and distant metastasis.[11] Therefore, it is important that radiologists be familiar with the imaging features of this pathology. We emphasize the need to consider phyllodes tumor as a differential diagnosis, mainly when solid masses with cystic areas are detected. The radiological suspicion for a phyllodes tumor already indicates surgery with wide excision, even if the confirmation on the basis of a needle biopsy is not possible, in order to avoid recurrence which may be malignant.

Largest phyllodestumour reported was of 50 x25.2 x16.4 cm in size[15], another was to be reported as 30x26x21 in size[16], another phyllodes tumour ever reviewed was about 24x 20 cm in size[13], another case report is of 20 cm size phyllodestumour[14]

Conclusion

The diagnosis of phyllodes tumor should be entertained in all patients presenting with progressive enlargement of breast lump. Emphasis should be given on proper health education on breast cancer and the negative consequences of alternative or delayed treatment. Excision should be done as soon as possible, as unnecessary delay can lead to disease progression and increased morbidity and mortality.

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