Fungating malignant phyllodes tumour- Still a reality in this day and age !!!

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Citation

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Abstract

Cystosarcoma phyllodes is an uncommon neoplasm of the breast, constituting 0.3–0.9% of all breast tumors. The incidence of malignant phyllodes tumor is even lower. The tumor is similar to fibroadenoma in structure, but it is distinguished from it histologically by large leaf-like projections of stroma with increased stromal cellularity[1]. Although surgical removal is the mainstay of treatment, the extent of surgery required (excision vs. mastectomy) and the need for additional local therapy, such as radiotherapy, are unclear[2] . We report a case of a 35 year old married woman who presented to our out patient department with a fungating, foul smelling mass in the right breast that turned out to be a malignant phyllodes tumour. The malignant variant of the cystosarcoma phllodes tumour is indeed a very rare mammary tumour.

INTRODUCTION

Cystosarcoma phyllodes is a rare, predominantly benign tumor that occurs almost exclusively in the female breast. Its name is derived from the Greek words sarcoma, meaning fleshy tumor, and phyllon, meaning 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 (hence the name). Because most tumors are benign, the name may be misleading. The World Health Organization in 1981 adopted the term, phyllodes tumour as this does not imply any biologic behavior, and it has gained wide acceptance[3]. Thus, the favored terminology is now phyllodes tumor, which is also used in the world health organization classification of breast tumours. To prevent confusion, the WHO classification should be strictly adhered to so as to standardize treatment results and to compare results from different centers. Thus phyllodes tumours may be malignant(25%), benign(60%), or indeterminate, the so called borderline lesion(15%). Phylloides tumours comprise less than 1% of all breast neoplasms and are unique in their occurrence exclusively in the female breast and appearance in no other site in the body.[4]

CASE REPORT

A 35 year old married nulliparous housewife presented to our out-patient department with a history of a lump in her right breast for a period of two months. The lump though initially small started to rapidly increase in size, and to become congested and heavy. The lump had burst open over the previous ten days with initial discharge of blood and then pus and extremely foul odour. She did not complain of fever, jaundice, abdominal lump or any bony pains.

On examination there was no palor or icterus, the pulse and blood pressure was normal. There was a foul smelling fungating mass measuring 20x18x16 cms almost replacing the right breast. The nipple areola complex was not seen and the central two thirds of the breast was replaced by a fungating tumoural mass, that exuded blood and pus and had a horrendous odour. The little skin left at the periphery of the mass revealed dilated veins in the subcutaneous plane. This skin was mobile over the tumour at the periphery. The whole tumoural mass and whatever little of the breast that was left, was mobile over the underlying chest wall. Axillary lymph nodes where palpable two in number largest 1x2cms and mobile. The left breast and axilla was normal. The respiratory and abdominal examinations revealed no abnormality.

Since the mass had already fungated we proceeded with an incisional biopsy, which was reported as malignant phyllodes tumour. All blood investigations, chest xray and abdominal ultrasound examinations where normal. A right mastectomy was done. We took a one centimeter margin from the clinically palpable periphery of the tumour, and after completing the mastectomy, we had a huge defect to

cover. We did not desect the axilla formally but removed low lying palpable lymph nodes. Keeping in mind the patients young age, we covered this defect with an extended composite latissimus dorsi musculocutaneous flap that provided soft tissue coverage as well as a breast mound that could later be contoured for symmetry, although cosmesis was not our primary aim. Post operatively the patient made a fairly uneventful recovery, and was discharged on the 15th post operative day. At discharge the patient was very happy and relieved, and thanked us for getting rid of the fungating foul smelling mass in her right breast. The final histopathology report was reported as high grade malignant phyllodes tumour, showing spindle shaped cells arranged in whorls and interlacing fashion, with cells showing moderate amount of cytoplasm and hyperchromatic spindle shaped nuclei, exhibiting moderate mitotic activity. Three axillary lymph nodes isolated revealed features of reactive hyperplasia. Apart from regular followup, we have advised the patient adjuvant radiotherapy because she had such a large tumour.

Figure 1

Figure1: fungating breast mass (three different views of the same mass.)

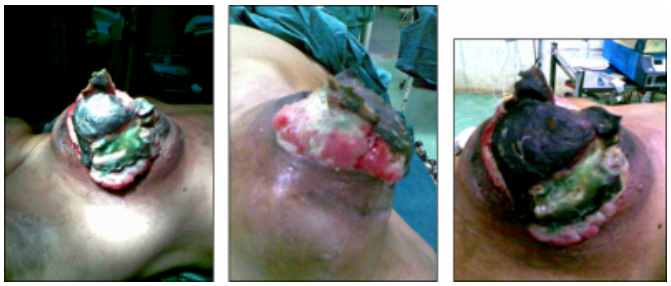


Figure 2

Figure2: post operative wound 3 weeks days after surgery showing the breast mound.

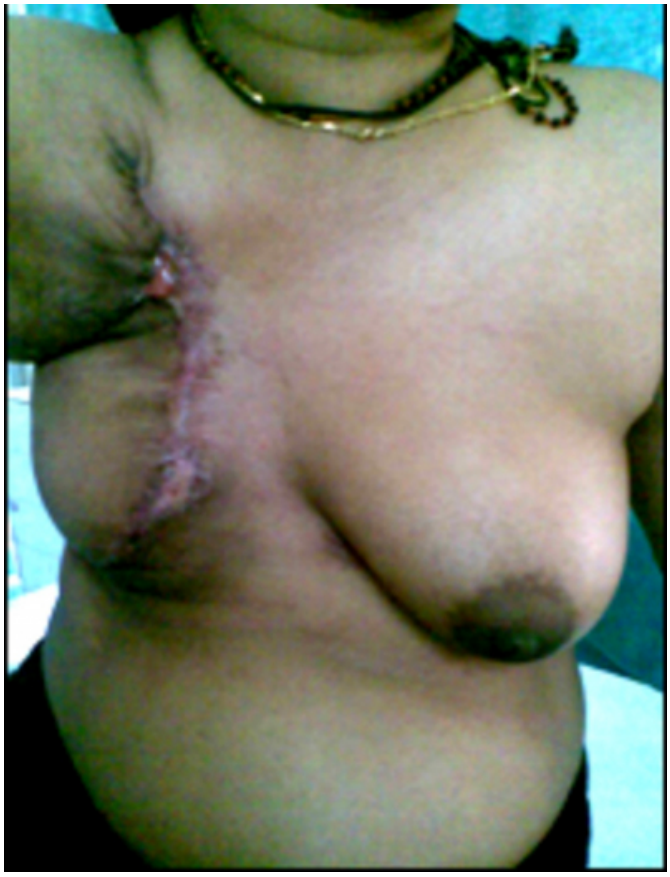
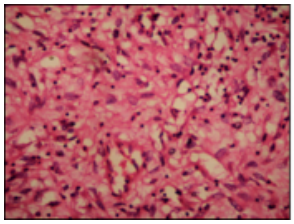
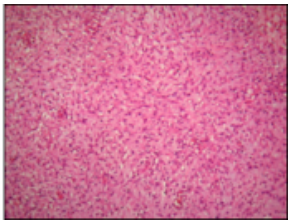


Figure 3

Figure 3: Histopathology slides with stromal cellularity, showing spindle cells with pleomorphism in a fibromyxoid background (high power x400, and low power x100.)





DISCUSSION

Phyllodes tumor is the most commonly occurring nonepithelial neoplasm of the breast, although it represents only about 1% of tumors in the breast[4] The tumor was first described by Muller in 1838[5]. Since then, numerous small and large series have appeared in the literature. There is still considerable uncertainty about the biological behavior of the disease. It has a smooth, sharply demarcated texture and typically is freely movable. It is a relatively large tumor, with an average size of 5 cm. However, lesions of more than

30 cm have been reported. Although the benign tumors do not metastasize, they have a tendency to grow aggressively and can recur locally.[2] Similar to other sarcomas, the malignant tumors metastasize hematogenously. Unfortunately, the pathologic appearance of a phyllodes tumor does not always predict the neoplasm's clinical behavior. In some cases, therefore, there is a degree of uncertainty about the lesion's classification. 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. Most patients with metastases die within 3 years of the initial treatment.[6]

Clinically a firm, mobile, well-circumscribed, nontender breast mass is appreciated. Curiously, cystosarcoma phyllodes tends to involve the left breast more commonly than the right one. Overlying skin may display a shiny appearance and be translucent enough to reveal underlying breast veins. Physical findings (ie, the occurrence of mobile masses with distinct borders) are similar to those of fibroadenoma.[7] Phyllodes tumors generally manifest as larger masses and display rapid growth. Mammographic findings reveal the appearance of round densities with smooth borders and are also similar to those of fibroadenoma. Recurrent malignant tumors seem to be more aggressive than the original tumor.

For benign lesions excision with a 1 cm minimum negative margin is advocated. For malignant tumours, if adequate margins are achieved with breast conserving surgery, mastectomy is not required. Formal axillary dissection seems to be unnecessary, but removal of low axillary lymph nodes cannot be criticized, especially if patients have palpable lymphadenopathy.[4]

Adjuvant radiotherapy may be considered for high risk phyllodes tumours, including those greater than 5cms, with stromal overgrowth, with more than 10 mitoses per high power field, or with positive margins.[8] RT is used as an adjuvant for acquiring maximum local control. RT after local wide excision is thought to decrease the local failure rates; however, it has been found to have no significant effect on survival[9]. There is no clear indication for adjuvant chemotherapy for patients with phyllodes tumours[10] However, its role is limited to the treatment of metastasis and for palliation of unresectable local recurrences.

Local recurrences are usually seen within the first few years of surgery, at the site of original excision, and are inversely proportional to the width of the negative resection margin. The recurrences are more aggressive histologically than the primary, and this condition is referred to as malignant transformation. Most patients with local recurrences are treated with total mastectomy.

Most metastatic pyhllodes tumours have spread hematogenously to lung, bone, abdominal viscera and mediastinum. Thre are no reports of long term survivors. The optimum palliative treatment of metastatic phyllodes tumour has not been found. Cyclophosphamide, doxorubicin, cisplatin and etoposide have been used with limited success. Radiation in symptomatic metastasis may be helpful. The majority of tumours contain estrogen or progesterone receptors, although palliation with hormone manipulation has not been extensively explored[4].

Malignant behavior is mainly decided on histopathology alone, as immunohistochemistry and cytometery are of no help [11]

CONCLUSION

Malignant phyllodes tumour is an extremely rare breast tumour. The WHO classification should as far as possible be adhered to, so as to prevent confusion and streamline results of various trials and studies. Stromal overgrowth, larger tumor size, and involved margins were all significantly correlating with local recurrences. Stromal overgrowth carries a grave prognosis .Breast-conserving surgery with appropriate margins is the preferred primary treatment. Mastectomy should be reserved for larger, more than 5cms lesions, high risk tumours and recurrent lesions. The current study data do not support the use of adjuvant radiotherapy for patients with adequately resected disease. Patients with stromal overgrowth, particularly when the tumor size was more than 5 cm, were found to have a high rate of distant failure, and such patients merit consideration of a trial that examines the efficacy of systemic therapy.

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