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Case Report

A rare case of giant malignant phyllodes tumour and long term survival

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ABSTRACT

Background: Phyllodes tumours are rare breast neoplasm. Its clinical spectrum ranges from a benign and locally recurrent form of behaviour to malignant and metastatic forms. The age at risk is between 35 and 45 years. Giant phyllodes tumours are larger than 10cm in diameter. **Case Report:** Herewith, we present a rare case of giant malignant phyllodes tumour (40x30x20cm) in a young lady who survived 10 years post-treatment in which most literature reported 10-year survival of 23% to 42%. The patient had mastectomy and radiotherapy to the anterior chest wall. **Conclusion**: A young lady with a giant malignant phyllodes tumour survived 10 years following treatment.

Keywords: *Long-term survival, Malignant giant phyllodes tumour, Young lady*

Introduction

Phyllodes tumour was first described by Johannes Muller in 1838 and has presented a diagnostic and treatment dilemma for physicians since its original descriptions.[1] Classically, the name cystosarcoma phyllodes was assigned because of the tumour's fleshy appearance and tendency to contain macroscopic cysts.[1]

Phyllodes tumour of the breast is a rare tumour type among all kinds of breast tumours, its incidence rate is 2% to 3% in all breast fibrous epithelial tumours or 0.3% to 1.0% in all breast tumours.[2] They are classified according to their biological behaviour: benign, malignant and borderline.[2] The existing diagnostic method has low diagnostic accuracy in general. Preoperative diagnosis uncertainty has hindered the rational development of surgical options.[3]
The first treatment of choice is surgical excision with negative surgical margins and is associated with relatively high disease-free survival and long-term survival rate and a low recurrence rate.[4] Radiotherapy is often used because phyllodes tumour tends to be locally aggressive.[4] The role of chemotherapy and hormonal therapy has not been established.[4]

Case report

A 28year old female student presented to our hospital with a mass in her right breast for 2 years. The patient had a lumpectomy in which histology revealed a benign phyllodes tumour. Recurrence was noticed a year later with multilobulated right breast mass, with accelerated growth in recent months. There was no family history of breast cancer.

Physical examination revealed a young lady, afebrile, pale, anicteric, not cyanosed, not dehydrated and no pedal oedema. The breast examinations showed a giant tumour of the right breast, with ulceration, exudates

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and bleeding. The other breast and both axillae were free. Other systems were essentially normal. Investigations revealed; full blood count (FBC) showed a packed cell volume (PCV) of 20% and white blood cell (WBC) of 14.0x 10^3/ul. Chest x-ray, abdominopelvic ultrasound and blood chemistries were all normal. The patient was commenced on antibiotics and also transfused with 4 pints of blood. The repeated FBC was normal.

The patient also had a simple mastectomy. The gross pathological specimen revealed a fungating huge right breast mass measuring about 46x31x22cm, 12kg in weight and a density of 0.820g/cm^3. Histology report revealed a section of breast tissue show a highly cellular stroma with tumour giant cells has hyperchromatic nuclei. There is a glandular lining at the top left corner. The postoperative course was uneventful and the patient was discharged after 4 weeks from surgery.

The patient also had external beam radiotherapy 30Gy in 10 fractions. The patient has been on regular follow-up and was seen with no evidence of recurrence. Chest and abdominal CT scans were normal.

Figure 1: Photograph of the lesion before surgical excision.

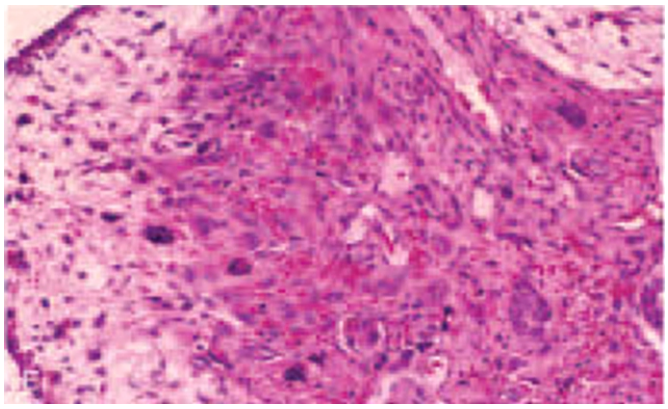

Figure 2: Malignant Phyllodes tumour shows highly cellular stromal with tumour giant cells, nuclear pleomorphism and mitotic activity of the stromal component (H&E, X400)

Discussion

Phyllodes tumour occurs predominantly in middle-aged or elderly women and its occurrence in younger patients as in the present case is exceptional.[4] The aetiology of this rare disease is still unknown. However, its incidence is higher in whites in general, in Latin whites and East Asians in particular.[5]

Patients usually present with a firm, rounded, mobile, well-defined and painless mass without any significant mammographic or ultrasound features.[1] Clinically it resembles a fibroadenoma and can be mistakenly left alone.[1] In most patients, axillary lymph nodes are not palpable at presentation, which is similar in this patient, because the metastatic spread of these tumours is primarily haematogenous (3% to 13%)5, the most common site is the lung.[6] The size of the tumour is variable, ranging from 1cm to >40cm.[7]

Making an early diagnosis of phyllodes tumour is extremely difficult. A variety of techniques, including colour Doppler ultrasound, magnetic resonance imaging (MRI), fine needle aspiration cytology (FNAC) and core tissue biopsy have been utilized to improve the pre-operative diagnosis.8[9,10]

We believe this case has one of the largest phyllodes tumours of the breast. In previous literature only three cases of large phyllodes tumour were reported; Kumar et al stated that their patient had the largest phyllodes breast tumour that measured 50x25.2x16.4cm and weighed 15kg with a density of 0.726g/cm^3. 11 Xia et al presented their case as a giant phyllodes tumour of the breast which

measured 47.5x37.0x28.0cm and weighed 9.79kg ex-vivo with 0.199g/cm^3 in density. 12 While sarvanandan et al presented their case that measured 28x25x18cm in size and weighed 8.27kg with a density of 656kg/cm^3.[13]

The treatment for phyllodes tumour remains surgical excision. However, for a very large tumour size like our patient, a total mastectomy is recommended to ensure complete removal. It is essential to keep a sufficient margin of healthy tissues to reduce the risk of local recurrence.[14] For borderline or malignant phyllodes tumours, or in cases of local tumour recurrence, mastectomy may be the preferred option.[14]

The role of adjuvant treatment with radiotherapy or chemotherapy 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 recurrence and distant metastasis.[14] Some authors advise radiotherapy which has been proven to improve patient survival and reduce local recurrence.[15] A consensus on chemotherapeutic protocols for this kind of neoplasm has not been achieved yet.[15] August and Kearney recommended that adjuvant radiotherapy be considered for high-risk patients, including those tumours>5cm, with stromal overgrowth, with > 10 mitoses/high power fields, or with infiltrating margins.[5]

Local recurrence is common in phyllodes tumours; 21% for benign types, 46% for borderline types and 65% for malignant types.[16] Prognosis is generally good, with 5-year survival rates of 91% and 82% for benign/borderlines types and malignant types respectively.[17]

Conclusion

Phyllodes tumours are rare breast neoplasm that presents as painless breast masses. Its prognosis and treatment are still debatable. Surgical excision with a clear margin remains the treatment of choice for these tumours. Herewith, reported a case of a giant malignant phyllodes tumour in a young lady who survived 10 years post-treatment. We believe this patient is among the largest phyllodes tumour reported in the previous publications.

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