

CASE REPORT: MASSIVE RUPTURED MALIGNANT PHYLLODES TUMOUR

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ABSTRACT

Phyllodes tumour is a rare entity, affecting mainly middle aged women. It consists of a spectrum of disease from benign tumour to highly aggressive malignant form. We present a case of massive ruptured malignant Phyllodes tumour, and its subsequent management.

Keywords: *Breast, palliative care, Phyllodes tumour*

Introduction

Phyllodes tumors, or cystosarcoma phyllodes, are fibroepithelial tumours of the breast. It is a rare entity, accounting for an estimated 0.3-0.9% of all breast tumours (1). Its incidence peaks at the 4th decade of life (1). Histologically, phyllodes tumours have both an epithelial component and mesenchymal (stromal) components, and can be classified into benign, borderline or malignant subtypes (2). Surgery remains the preferred modality of treatment in patients with malignant phyllodes tumour and complete R0 resection offers high rates of local control (3). We present a case of a massive right breast sarcoma with rib and pleural metastasis and its subsequent management, which required staged surgeries that ultimately resulted in improved quality of life.

Case report

A 58-year-old Chinese lady presented to our Plastic Surgery clinic with a complaint of a right breast lump for the past one year. Despite being aware of it, she did not seek medical treatment till the tumour impedes her movements, ruptured and became unbearably foul smelling for the past two months. Her right arm is kept in a fixed, abducted position with minimal mobility of the right shoulder as a result of the massive breast tumour. She is menopausal for 10 years, married with one child. Family history was unremarkable, and there was no history of psychiatric illnesses. On examination, she appeared severely cachectic,

with a body mass index of 16.3 kg/m². There was a huge fungating right breast tumor extending from the clavicle to the umbilicus level, with extensive skin involvement till the right nipple is not discernable. Multiple areas of foul smelling pus discharge were also seen (Figures 1a and 1b). The tumour is also fixed to the chest wall. There were no clinically palpable lymph nodes. Computed tomography (CT) of the thorax and abdomen revealed a large tumour (Figure 2b) with extension to the rib and pleura. Her core biopsy result was inconclusive.

She underwent a right toilet mastectomy with the aim of palliative care. The tumour was dissected off the right 6th rib en bloc with the pectoralis major muscle (Figure 2a). The underlying wound was too large to be closed primarily. Histopathological examination revealed a 6.0 kg malignant phyllodes tumour measuring 40 cm x 38 cm x 16 cm. All margins were involved, Ki67 proliferation index was 20-30%. Post-operative positron emission tomography (PET) scan noted tumour infiltrating the 5th and 6th ribs, but no distant site uptake. After one month of admission for wound management, physiotherapy and nutritional support, she was discharged home with a much-improved quality of life. She was able to ambulate, carry out simple chores, was independent in her daily activities and her family were able to accompany her without the foul smelling fungating tumour. Six weeks later, she was readmitted due to pleural effusion and subsequently expired due to pulmonary complications.



Figure 1a and 1b: Lateral and anterior views of the large fungating right breast tumour

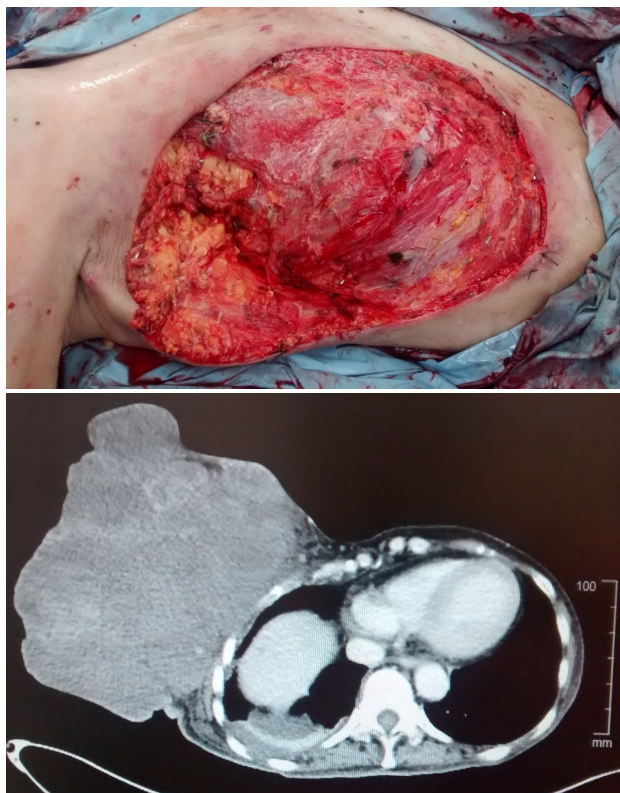


Figure 2a: Right chest wall wound after toilet mastectomy

Figure 2b: Selected Computed Tomography image showing a large tumor with rib and pleural involvement (blue arrow)

Discussion

Phyllodes tumours are rare breast tumours, which classically presents as a painless, firm, mobile and multiloculated breast lump which rapidly expands. Tumour rupture (4), nipple discharge and skin ulcerations are rare presentations (5). Axillary lymph nodes are only palpable in 20% of cases, but lymph node involvement of phyllodes tumor is rarely seen. Histologically, phyllodes tumours is characterised by epithelial lined cleft-like spaces with a hypercellular stroma, organised into leaf-like fronds. World Health Organization further classifies Phyllodes tumour into benign, intermediate, or malignant tumour based on the tumour margins, stroma cellularity, mitotic rate and pleomorphism. The lungs are the most common site for tumour metastasis. Liver, bone and distant lymph nodes are the other frequent sites of metastasis (6). The overall rate of all phyllodes tumour metastasis is reported as 4% (7).

Currently, there are no gold standard investigation for phyllodes tumour, as ultrasonographic and mammographic images have low sensitivity for differentiating phyllodes tumours and fibroadenomas (8).

The standard treatment for phyllodes tumour is a wide excision with margins of normal tissue greater than 1.0 cm. In cases of high tumour-to-breast ratios or recurrent tumours, mastectomy is recommended (9). There are still controversies with regards to adjuvant chemotherapy and radiotherapy in the post-operative management of malignant phyllodes tumours and is only used on selected cases after multidisciplinary assessments. In our case, a curative resection would include mastectomy, multiple rib resections and plurectomy. However, the patient was not optimised for such an extensive resection, and was only fit for palliative resection with the aim of quality of life improvements.

Most surgeons would not consider performing palliative surgery on such patients because the surgery would not change their prognosis or survival rate. Nevertheless, such palliative therapies would improve the patient’s psychosocial well-being, provide dignity and encourage family support in their short remaining life.

In conclusion, malignant phyllodes tumours, being rare fibroepithelial breast tumours, warrant the clinician to maintain a high index of suspicion if a patient presents with a rapidly enlarging breast lump. A simple palliative mastectomy should be performed if negative margins are not achievable by wide local excision to improve their quality of life. Adjuvant therapy is considered in individual patients.

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