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Malignant phyllodes tumour presenting as a massive fungating breast mass and silent thrombo-embolism

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ABSTRACT

INTRODUCTION: We report an unusual case of a massive malignant phyllodes tumour that had almost replaced the entire breast presenting with severe chronic blood loss, extensive deep venous thrombosis (DVT) and a silent pulmonary embolus.
PRESENTATION: Long-standing neglected massive fungating ulcerative mass larger than the left haemothorax.
DISCUSSION: Phyllodes tumours are rare fibro-epithelial breast lesions that have the propensity to grow rapidly to a large size if neglected. Larger tumours are more likely to be malignant with an overall metastatic rate around 10%. An incidental pulmonary embolus arising from extensive silent lower limb deep vein thrombosis requiring an IVC filter complicated the surgical management.
CONCLUSION: Phyllodes tumours are rare and account for approximately 0.3–0.5% of all breast tumours [1]. They have the propensity to be fast growing. However, tumours reaching a massive size (>10 cm) are rare with few reports in the literature.

1. Summary

Phyllodes tumours are rare and account for approximately 0.3–0.5% of all breast tumours [1]. They have the propensity to be fast growing. However, tumours reaching a massive size (>10 cm) are rare with few reports in the literature. We report an unusual case of a massive malignant phyllodes tumour that had almost replaced the entire breast presenting with severe chronic blood loss, extensive deep venous thrombosis (DVT) and a silent pulmonary embolus.

2. Background

This patient presented with a long-standing neglected phyllodes tumour that had grown to a massive fungating ulcerative mass larger than her left haemothorax. Investigations showed severe anaemia, lower limb DVT and a silent pulmonary embolus.

3. Case presentation

A previously healthy woman in her 40s presented to the emergency department with a massive exophytic left breast tumour (Fig. 1a). She successfully concealed this enlarging lump for 3 years (Fig. 1a). The patient was anaemic, with haemoglobin of 42 g/L, due to significant chronic blood loss from the tumour. An urgent incisional biopsy diagnosed a phyllodes tumour. CT confirmed the absence of metastatic spread. However, an incidental right lower lobe pulmonary embolus and extensive thrombus were seen within the right common femoral vein extending to the iliac vein. Due to the ooze from the fungating breast tumour she was unable to be anticoagulated and an urgent IVC filter was inserted.

4. Investigations

4.1. Radiology

Ultrasound (GE Logic E9 system, GE healthcare, UK) was performed as mammogram was not possible due to the size and nature of the tumour. This demonstrated a large left breast heterogeneous mass with marked posterior attenuation (Fig. 2a and b). On B-mode imaging, the internal architecture exhibited a macro-foliated echo-pattern intermixed with disorganised echogenic curvilinear foci

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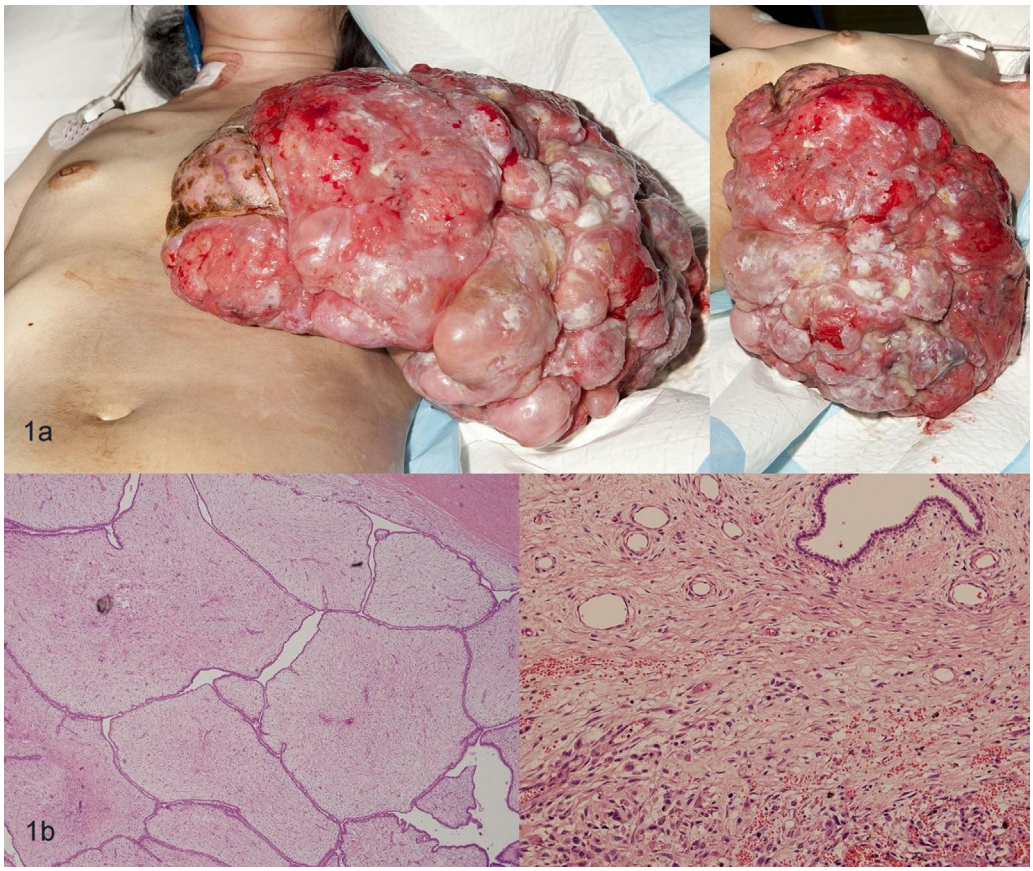

Fig. 1. (a) The ulcerated left breast mass, (b) low and high power histology.

on a heterogenous hypoechoic background. Superficially this was encapsulated in a rind of hyperechoic tissue. Colour Doppler study showed increased vascularity within the tumour. The deep margin was not assessable due to marked beam attenuation and depth of lesion.

Coronal CT image showed a massive left breast mass with superficial chest wall muscle invasion but no bony involvement. Parasitic vascular supply was predominately via large vessels arising from the left axillary and left internal thoracic arteries (Fig. 2c).

MRI was performed to further aid characterisation, delineate the deep margin and assess chest wall involvement. Multiplanar pre and post contrast acquisitions (Siemens Magnetom Aera 1.5T magnet, Siemens Healthcare, Malvern, PA), showed the mass had replaced the entire breast, with a maximal cranio-caudal length of 300 mm (Fig. 2d and e). The mass showed a massively exophytic growth pattern in an antero- lateral direction with a crenellated macro-foliated bright T2 (Fig. 2d and e) and iso- to low signal T1 (Fig. 2f) signal pattern. On the post contrast fat suppressed T1 study, the tumour exhibited encapsulated hypointensity posteriorly with a peripheral frill of hyperintense crenellated flanges extending anteriorly. Extension into the left axilla was noted. Separate to the tumour, left axillary lymphadenopathy was observed. Superficially the tumour exhibited a disorganised lobulated contour, at the deep margin there was superficial chest wall muscle invasion and parasitic vascular supply was noted. Importantly, no bony involvement was present.

Post contrast acquisitions (Fig. 2c) of the chest, abdomen and pelvis, as well as pre and post-contrast cranial CT (64 slice GE lightspeed VCT, GE healthcare, Little Chalfont, UK) did not reveal any distant metastatic focus. However, it did detect an incidental right pulmonary embolism and extensive right deep vein thrombosis.

4.2. Pathology

Macroscopic examination revealed the tumour to involve the majority of the breast tissue measuring 395 mm in maximal extent including a large exophytic ulcerated component. The nipple was preserved in the skin inferior to the tumour. On the cut surface, the tumour was circumscribed and lobulated with myxoid and firm pale fibrous areas. The tumour involved the deep margin of the breast over a broad front.

Histopathology showed a biphasic tumour with epithelial and mesenchymal components. The tumour had an intracanalicular growth pattern with cleft-like spaces and the epithelial component did not show atypia. The majority of the stromal component showed mild atypia and low mitotic activity. Focal areas of hypercellular stroma with moderate nuclear atypia and up to 12 mitoses per 10 high power fields were identified amounting to a malignant phyllodes tumour. In addition, a 10 mm focus of sarcomatous change with pleomorphic bizarre spindled cells and frequent mitoses was identified.

5. Treatment

The patient was given 3 units blood transfusion and had an IVC filter inserted prior to breast surgery. Left total mastectomy was performed with removal of underlying pectoralis major muscle. There was no involvement of intercostal muscles or ribs. The tumour measured $395 \times 245 \times 130$ mm. The resulting large skin defect was managed with a VAC dressing. The patient made good recovery post-operatively and was discharged 4 days later.