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

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ARTICLE INFO

Article history:
Received 28 January 2015
Accepted 17 February 2015
Available online 19 February 2015

Keywords:
Breast
Malignant
Tumour
Thromboembolism
Phyllodes

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.

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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 echopattern intermixed with disorganised echogenic curvilinear foci.
on a heterogenous hypoechoic background. Superficially this was encapsulated in a rim 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. Separately 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 × 245 × 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.
Fig. 2. (a and b) Ultrasound shows a large heterogenous mass with marked posterior attenuation. (c) Coronal CT shows parasitic vessels and exophytic nature of the mass. (d) Coronal T2, shows heterogenous intensity with a crenellated macrofoliated margin. (e and f) T1 pre and post contrast images, show an enhancing rim.

6. Outcome and follow-up

Further resection of positive margin was performed one month later. At four months follow-up, the wound was granulating well with no skin grafting required for closure. The first annual follow up showed no evidence of recurrence. However, subsequent surveillance imaging and biopsy of her contralateral breast showed widespread ductal carcinoma in-situ for which she still refuses surgery.

7. Discussion

Phyllodes tumours are rare fibroepithelial breast lesions that have the propensity to grow rapidly to a large size if neglected. They are classified histologically as benign, borderline or malignant with larger lesions more likely to be malignant. This is suggested by a single institution study of 65 cases which found that the mean volume of tumour was higher in the malignant group [2]. The World Health Organisation reports an overall metastatic rate of 10% with most arising from the malignant category [3]. Staging investigations in our patient detected an incidental pulmonary embolus arising from lower limb deep vein thrombosis. Due to the large oozing ulcerative surface of the tumour, she went on to have an IVC filter preoperatively and subsequent postoperative anticoagulation. While the incidence of venous thromboembolism in breast cancer patients is reported to be 1.2% [4], the incidence in patients with malignant phyllodes is unknown. It may be prudent to consider the diagnosis and if necessary exclude this in patients with massive phyllodes tumours that undergo preoperative staging given their higher chance of malignancy.

The risk of tumour recurrence is strongly related to the excision margins. A recurrence rate of 15–20% is reported for tumours with margins of <1 cm [5]. However, in the absence of an adequate surgical margin, localised radiotherapy may be considered. A recent prospective study reported no local recurrence at a mean 56 month follow-up in 46 patients treated with this strategy [6].

Conflicts of interest

All authors declare that they have no financial and personal relationships with any other people or organisations that could inappropriately influence (bias) this work including by employment, consultancies, stock ownership, honoraria, paid expert testimony, patent applications/registrations, and grants or other funding.

Funding

There was no source of funding for this work. All work was undertaken by the authors in their own time.

Ethical approval

Approval has been obtained from HREC SCGH (ethics committee). Reference = QIA # 4133.
Author contribution

This manuscript has six contributing authors who have all contributed to the case report, literature search and write up of the case. All authors had access to the patient information. All authors have read and approved the final manuscript.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Declaration

I, Anita Geraldine Bourke, the corresponding author, has the right to assign on behalf of all authors and does assign on behalf of all authors, a full assignment of all intellectual property rights for all content within the submitted case report in any media known now or created in the future, and permits this case report (if accepted) to be published in the International Journal of Surgery Case Reports.

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