CASE REPORT

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A giant rapidly progressive breast phyllodes tumour causing a skin rupture. A case report and literature review

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ABSTRACT

Large phyllodes tumours may behave in a rare way, in this paper we present a case of large phyllodes tumour with a female lady aged 42. The tumour caused a skin rupture due to rapid growth.

Keywords: breast phyllodes, mammogram, mastectomy.

Introduction

1% of all known breast neoplasms are phyllodes. In 1838, Johannes Muller of Germany suggested the term cystosarcoma phyllodes to describe them. Breast phyllodes tumour (BPT) is a tumour of fibro-epithelial cells with classically deep "leaf-like" projections into cystic spaces and sarcomatous stroma[1]. As 70% of theses lesions are benign, and only rarely demonstrate cystic features. World Health Organization (WHO) currently recognised the term Phyllodes tumour as the most appropriate nomenclature. The tumour size is variable, ranging from 1 cm to 50 cm [2]. The histological classification is determined by different elements including stromal atypia, stromal overgrowth, mitotic count and tumour margins. After the analysis of these criteria the tumour will be given a grade from a spectrum of benign, borderline, borderline/malignant and malignant features [3]. This is a paper reporting a very large phyllodes tumour with unusual skin manifestations, due to large tumour size, rapid growth, high body mass index (BMI) and large breast size the patient developed skin rupture.

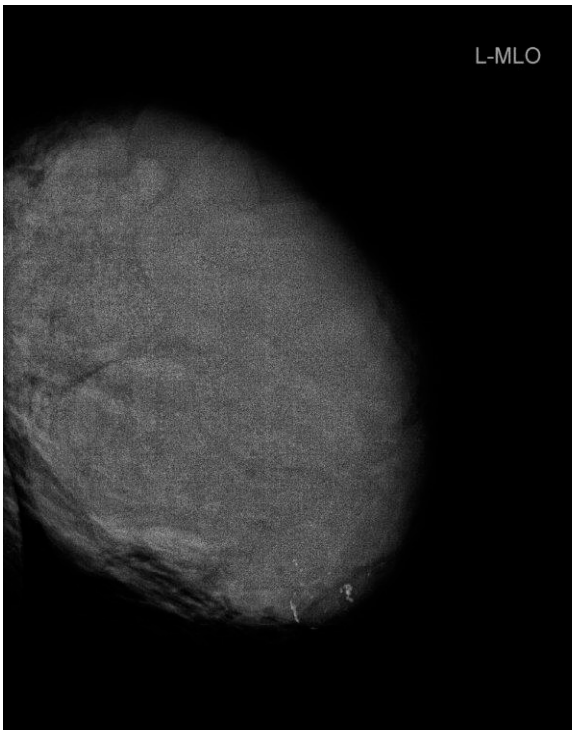

Figure 1. Mammogram revealed diffuse density of the whole breast

Case report

A 42-year-old previous fit female patient presented a two weeks history of palpable mass on the left breast, with accelerated growth pattern. There was no personal or family history of breast or ovarian cancer. Clinically in spite the patient has high BMI, there was obvious breast asymmetry the whole left breast was occupied by large lobulated firm mass and stretching the skin over. There were no signs of axillary lymphadenopathy.

Breast imaging with mammogram (**Figure 1**), breast ultrasound and CT scan (**Figure 2**) reported, a large lesion with internal vascularity and a heterogeneous echo texture is seen encompassing the entire left breast. In the axillary tail adjacent to the mass there are two cystic areas. Core biopsy was taken, the histology showed borderline phyllodes pathology. The patient in

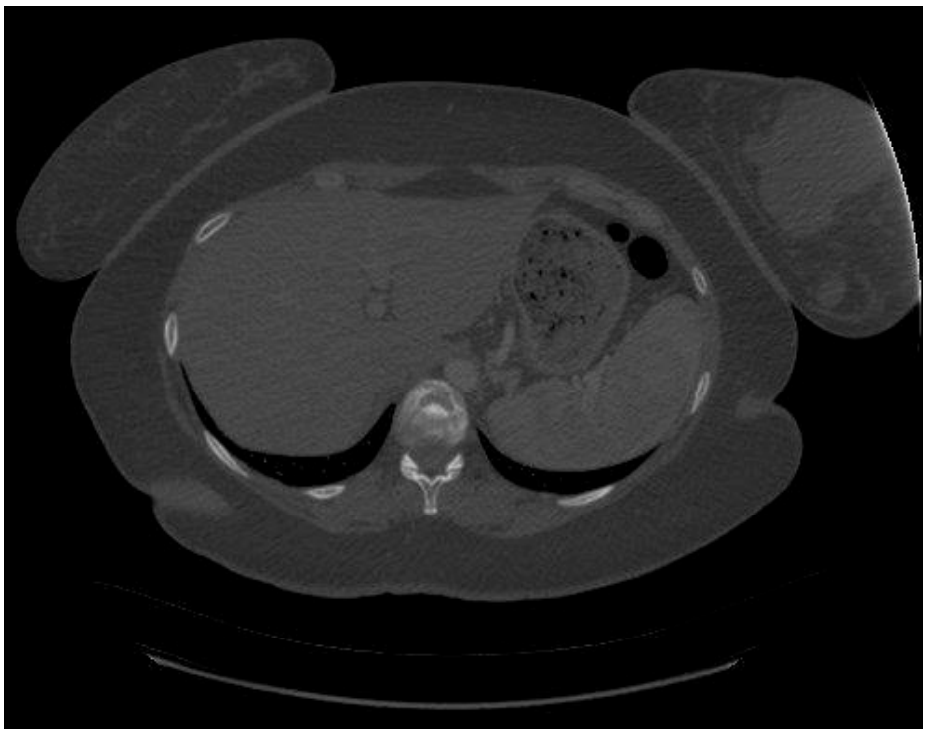

Figure 2. CT scan showing a large lobulated soft tissue mass in the left breast

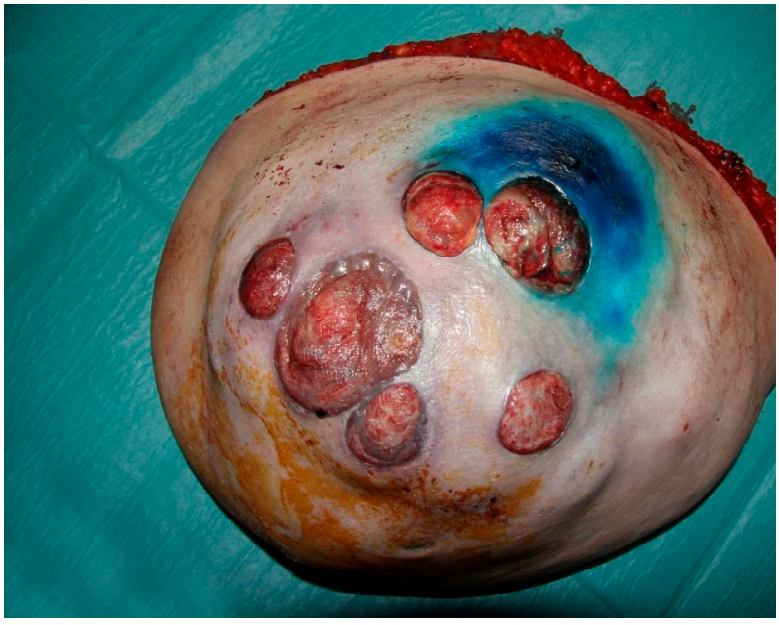

Figure 3. Mastectomy specimen with tumour protrusion through skin rupture

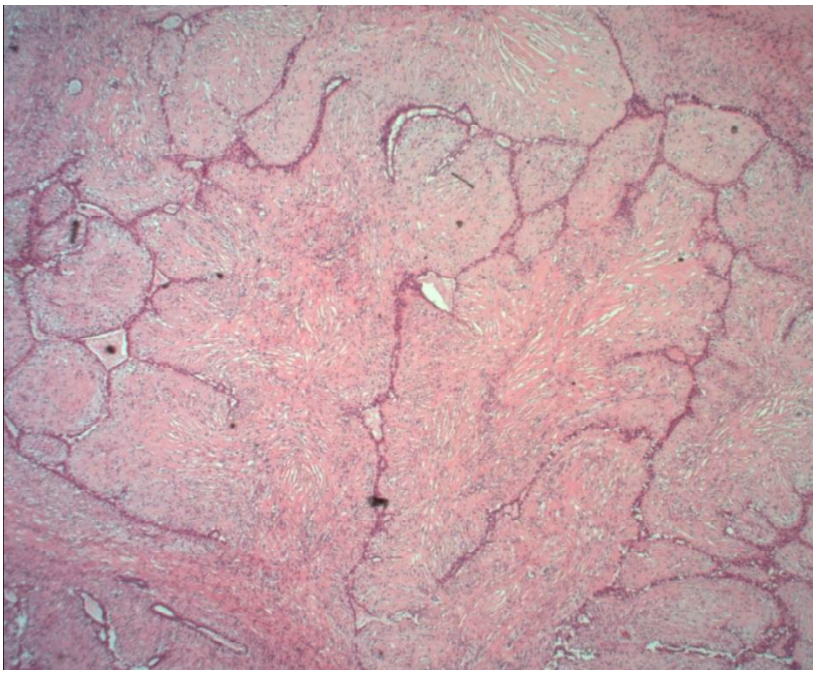

Figure 4. Low grade phyllodes displaying complex epithelial arrangement, cleft-like spaces and concentration of tumour cells beneath the epithelium

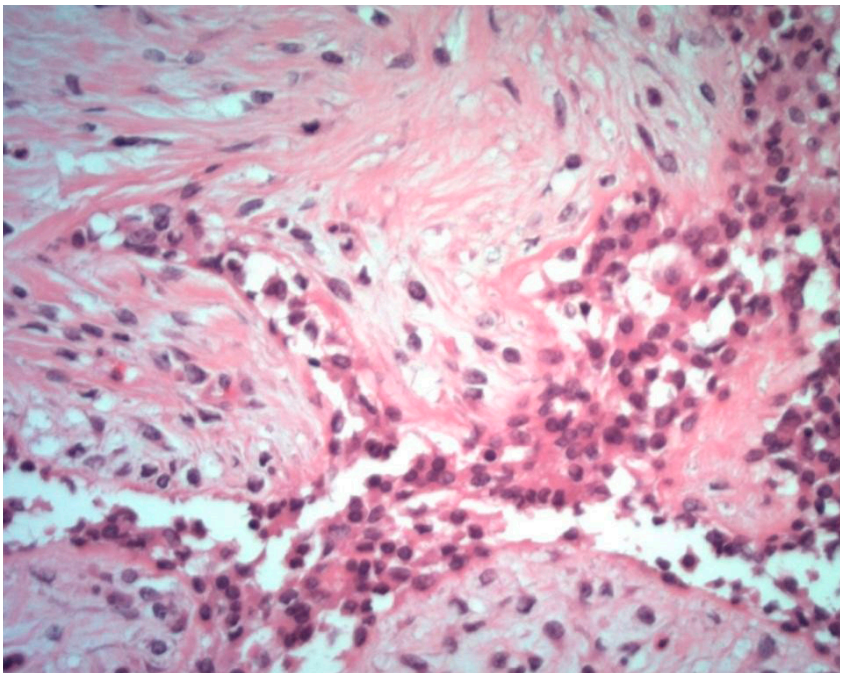

Figure 5. Low grade phyllodes with single mitotic figure in tumour cells beneath the epithelium

few days developed skin rupture ulcerations and the tumour protruded through the skin defect. An expeditious surgical management with mastectomy and sentinel lymph node biopsy performed after discussion in multidisciplinary team meeting. The postoperative histology showed fibro-adenomatous growth pattern with a cellular stroma component. There is a surface ulceration and areas exhibiting a leaf like growth pattern. This confirmed the pre-operative diagnosis of border line phyllodes pathology (**Figures 4, 5**).

Discussion

The phyllodes tumor was first described in 1827 by Chelius [4], however in 1838, Johannes Muller was the first one who applied the name cystosarcoma phyl-

lodes, and described it as a huge neoplasia with a cystic lobulated section and rapid growth [5].Initially it has been attracted as totally benign, but Cooper and Ackerman, in the year 1943 reported on the potential malignant biological pattern of this tumour [6]. Breast phyllodes peak incidence between 35–55 years of age in white females [7, 8], also prevalent in Latin American, and Asian populations [6]. Usually are unilateral, single, nodular, painless masses with an insidious onset and slow progression [9] with size may grow up to 50 cm in neglected cases [6, 10]. A rare features as skin rupture, may be related to rapid growth of the lesion. Wijeyaratne in 2010 and Nabi et al, in 2013 reported a cases of breast phyllodes causing breast skin rupture [11, 12]. As as in our case, probably the obesity and high volume of breast masked the breast tumor growth and contributed to skin rupture.

Phyllodes tumours of the breast arises from the fibroepithelial component of the breast and are rare in general and their incidence ranges from 0.3–1% of all breast tuomors [13], also they form a rate of 2% to 3% in all breast fibrous epithelial tumors. Rowell in 1993, reported the incidence is about 2.1 in one million [14]. They are characterised by local recurrence which the most important prognostic feature of this condition where the nodal or distant metastases are rare. The local recurrence incidence rate is about 15%, it is has been reported that, the incomplete excision is a contributary factor to the recurrence [13]. Borderline and malignant phyllodes tumor metastasis rate is about 25% to 31%, however The overall rate of all phyllodes tumor metastasis being 4% [15, 16]. The modes of tumor metastasis are primarily via blood, rarely lymph nodes. Chest wall soft tissue was the most frequently affected site of distant metastases, followed by the lungs, thoracic cavity, bones, and pleura. A rarer sites for metastatic lesions as vulva also has reported [17]. Chest wall soft tissue lesions may be related to incomplete local tumour excision and most patients with distant metastasis had progressed from local recurrence. The predictive factors for local recurrence were age, tumor size, histotype, and margin status. It is generally advised that the resection margin distance should be at least 1 cm for adequate surgical resection [18]. In addition to local recurrence, other risk factor for distant metastases are stromal overgrowth [19] and the tumour histological subtype. Kim et al. reported that the expression of stromal matrix metalloprotein-14 (MMP-14) was associated with a higher rate of recurrence [20]. There is a clinical diagnostic dilemma, as the history and clinical findings often mimic that of the common benign presentation of fibroadenoma or even cancerous lesions in rare occasions. The importance of phyllodes tumours today lies in the need to differentiate them from other benign breast lesions. The most common presentation is a solitary and unilateral breast lump, as do fibroadenomas. There are no pathognomonic radiological features could be detected in the mammogram or breast ultrasound for phyllodes tumour [21].Features that may indicate phyllodes diagnosis are: a large mass, 3rd and 5th decade of life, an increase in size of a previously stable mass and a rapid growth pattern. Other non-specific features as dilated skin veins, blue skin discolouration and nipple retraction [8]. This rare presentation should raise strong suspicion for Phyllodes. This is essential to differentiate it from fibroadenomas to avoid incomplete excision and local recurrence, also to differentiate it from breast cancer to avoid over treatment. Malignant transformation of a phyllodes tumour is a rare form of breast cancer, accounting for just 0.1–3% of all breast cancer cases [3, 8].

The tumour rarely shows cutaneous manifestations which may be confused with a cancer fungation. In this case the fast growth of the tumour exceeds the skin ability to stretch. This results in skin rupture and tumour protrusion through the skin defects. Also the tumour had expanded so rapidly that bleeding, infarction, necrosis and degenerative changes result in cystic lesions [22].

The histopathological morphology of the disease presents a continuum from benign to malignant nature. In 1981, the World Health Organisation (WHO), adopted the term phyllodes tumour. WHO classification of the phyllodes tumour into benign, borderline and malignant phyllodes, is based on amalgamation of various histopathological characteristics, including stromal cellularity, nuclear atypia, mitotic activity per high power field (HPF), stromal overgrowth, and tumor margin appearance [23, 24], see **Table 1**.

Surgery has been agreed the gold standard in the management of the phyllodes tumours ever since they were first described. Breast conservation surgery for small tumours and reasonable breast size. A clear margin of 1 cm is required to improve the outcome and reduce the risk of disease recurrence. Mastectomy is advisable in large tumours or small breast size. Immediate or delayed reconstruction still can be offered for mastectomy Adequate surgical excision with clear margins is the most essential preventive measures to minimize the disease recurrence. Using adjuvant treatment as radiotherapy or chemotherapy is debetable,

Table 1. Main histological features of the 3 grading subgroups for phyllodes tumours. Tavassoli FA, Devilee P. World Health Organization Classification of Tumours. 2003. HPF: High Power Field

	Benign	Borderline	Malignant
Stromal hypercellularity	Modest	Modest	Marked
Cellular pleomorphism	Little	Moderate	Marked
Mitosis	Few if any	Intermediate	Numerous (more than 10per 10 HPF
Margins	Well circumscribed pushing	Intermediate	Invasive
Stromal pattern	Uniform stromal distribution	Heterogeneous stromal expansion	Marked stromal overgrowth
Heterologous stromal differentiation	Rare	Rare	Not uncommon
Overall average distribution	60%	20%	20%

however some authors advise to use them in cases of malignant phyllodes [25, 26].

Conclusion

The patterns of phyllodes tumour are widely varied, making the clinical diagnosis. There should be a high index of suspicion for this diagnosis if there is a rapid progression of the tumour. It may causes rupture of the breast skin and presents as an external fungating breast mass, a presentation which is exceedingly rare. High body mass index or large breast size may mask the large tumour size and skin rupture may happens also. In spite it is a benign entity in most of the cases, an adequate clear surgical margins are very essential to minimise the recurrence and radical tumour removal remains the most crucial preventive measure against disease recurrence. Simple mastectomy is advisable if breast conservation will not secure a clear margin. Nodal metastases are rare and routine auxiliary dissection is not recommended.

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Conflict of interest statement
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