

Giant Phyllodes Tumor: A Case Report and Review of the Literature?

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ABSTRACT

Phyllodes tumor is a rare, fast-growing, fibroepithelial neoplasm of the breast that can be classified as benign, borderline or malignant. The case reported here refers to a 25-year old patient with a lump on her left breast that grew rapidly in the postpartum period, reaching 21 cm in size. Core biopsy and immunohistochemical analysis indicated a diagnosis of high-grade phyllodes tumor. The patient underwent mastectomy of the left breast followed by breast reconstruction. Histopathology confirmed a malignant phyllodes tumor with chondrosarcomatous and fibrosarcomatous differentiation and no angiolymphatic or perineural invasion. Phyllodes tumors are generally benign and differential diagnosis is usually made with fibroadenomas; however, clinical differentiation can be difficult in some cases. The definitive diagnosis is achieved by histopathology, which is able to differentiate benign from malignant forms of the disease. Standard treatment consists of local excision, with mastectomy being necessary in cases of borderline, large, malignant or recurrent tumors.

Keywords

Breast, Breast neoplasms, Mammoplasty, Mastectomy, Phyllodes tumor.

Introduction

Phyllodes tumors, also known as cystosarcoma phyllodes due to the presence of leaf-like projections of the tumor tissue extending into cystic spaces, is a rare type of fibroepithelial breast tumor that originates in the intralobular stroma. In the majority of cases they are benign and behave much like a fibroadenoma. The peculiar characteristic of these tumors compared to fibroadenomas is their increased stromal cellularity; hence they are also referred to as hypercellular fibroadenomas [1]. Phyllodes tumors are relatively

rare, corresponding to approximately 0.5% of all breast neoplasms, and their growth depends on certain factors including trauma, breastfeeding and pregnancy [2]. The tumors are characterized by rapid growth and can reach considerable sizes. They are more likely to affect women of 35-55 years of age and can be classified as benign, borderline or malignant [3]. The clinical case reported here concerns a rare malignant, rapidly growing phyllodes tumor that reached 21 cm in size, weighing 4,145 grams.

The institute's internal review board approved this case report under reference number 4.448.081. The patient gave her consent to publish these data.

Case Report

A 25-year old woman sought medical care at the breast outpatient clinic of the Santa Casa de Misericórdia Hospital in Vitória, Espírito Santo, Brazil, complaining of a painless lump on her left breast that had been present for ten years but that had increased significantly in size over the preceding eight months since the birth of her last child. Physical examination revealed an irregular, hardened, voluminous mass of approximately 20 cm that was painless at palpation and associated with localized warmth. Increased vascularity over the entire left breast was clearly evident (Figure 1). Lymph nodes were not palpable. Breast ultrasonography performed a month previously showed a heterogeneous multinodular mass in the upper inner quadrant of the left breast, measuring 15 cm and encompassing almost the entire breast. The mass contained oval anechoic areas and had irregular contours. Doppler revealed hyper vascularity.



Figure 1: Physical examination showing the presence of an irregular, hardened, voluminous mass measuring approximately 20 cm.

Core biopsy results suggested a high-grade (borderline or malignant) phyllodes tumor. Immunohistochemistry showed positivity for CD34 (stromal component), pan-cytokeratin (clone AE1/AE3) (epithelial component), c-Kit (CD117) proto-oncogene (clone EP10) (epithelial component), and Ki-67 (around 15% of the neoplastic cells in the stromal component), raising suspicion of a high-grade, borderline phyllodes tumor.

The patient underwent mastectomy of the left breast followed by breast reconstruction (Figure 2). A tumorous mass measuring 21 x 19 x 15 cm and weighing 4,145 grams was removed (Figure 3). Histopathology revealed a malignant phyllodes tumor with chondrosarcomatous and fibrosarcomatous differentiation and no angiolymphatic or perineural invasion (Figures 4, 5 and 6).

Computed tomography scans of the brain, chest and abdomen were negative for distant metastases. The patient was submitted to adjuvant chemotherapy due to the aggressive aspect of the disease. The treatment proceeded without complications.



Figure 2: Cosmetic outcome of immediate post-mastectomy reconstruction of the left breast.

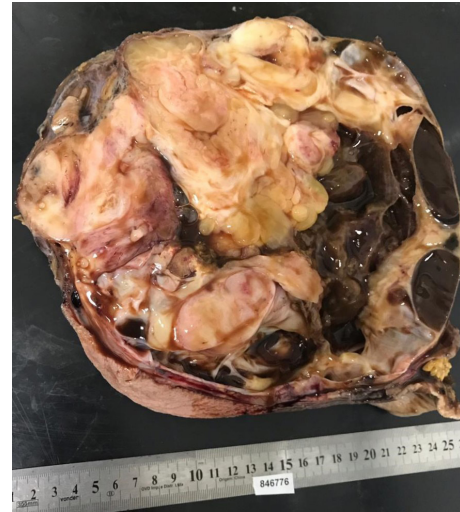


Figure 3: Macroscopy: a fleshy-looking solid mass containing peripheral cystic areas (dimensions: 21x19x15 cm; weight: 4,145 grams).

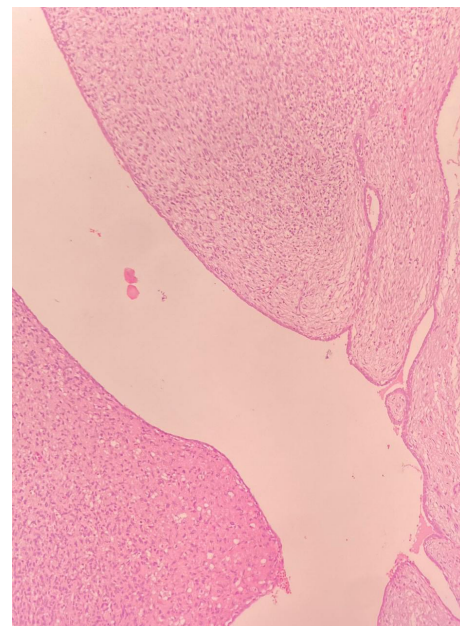


Figure 4: Biphasic epithelial and stromal proliferation showing high stromal cellularity and leaf-like projections (hematoxylin-eosin, 200X).

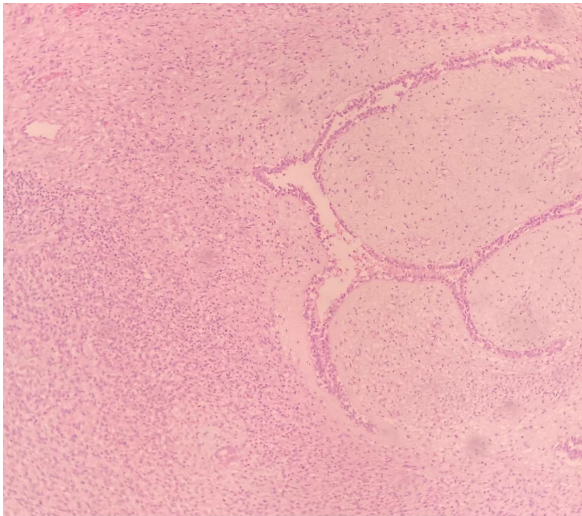


Figure 5: Histological pattern showing biphasic proliferation with increased stromal cellularity (hematoxylin-eosin, 200X).

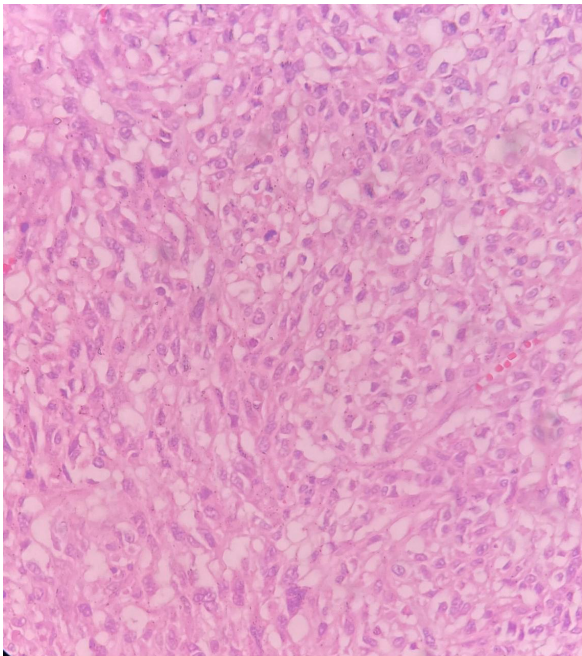


Figure 6: Histological features: marked atypia and high mitotic index (hematoxylin-eosin, 400X).

Discussion

Phyllodes tumors or hyper cellular fibroadenomas account for 2-3% of all fibroepithelial neoplasms of the breast [4] and 0.3 to 0.9% of all breast tumors [5]. These rare tumors are generally benign and differential diagnosis is made with fibroadenomas, although in some cases clinical differentiation can be difficult. However, peak incidence is around ten years later in the case of phyllodes tumors compared to fibroadenomas [6,7]. Overall, 58.4% to 74.6% of all phyllodes tumors are benign, while 15.0% to 16.1% are borderline and 9.3% to 31% are malignant [5]. They normally present as a palpable, elastic, moveable mass that is lobulated and painless.

Although size may vary, mean size tends to be around 5 cm and there is generally a history of recent rapid growth. Malignant phyllodes tumors tend to progress rapidly and are aggressive, whereas benign tumors are smaller and slower growing [3].

Diagnosis is clinical, and mammography is unnecessary; however, definitive diagnosis requires histopathology, which is also able to differentiate the benign from the malignant forms. The accuracy of fine-needle aspiration is 23%, while that of core biopsy is 65% [4]. No predisposing factors have yet been defined and the etiology of the phyllodes tumor remains to be fully clarified. The presence of estrogen and progesterone receptors in the phyllodes tumor suggests that hormonal factors may affect its growth [8]. Some factors such as lactation and pregnancy appear to accelerate growth, which may explain the rapid growth and large volume of the tumor found both in the case reported here during the postpartum period and in that reported by Carvalho Junior et al. [2] during pregnancy.

Prognosis is generally good; however, recurrence is a common setback, occurring in 15-17% of cases, particularly in the first three years [3]. Surgery is the standard treatment, with local excision in cases of small, benign tumors, leaving a margin of 1-2 cm to reduce the risk of recurrence. In cases of borderline, large, malignant or recurrent tumors, total mastectomy is necessary. Lymph node involvement is infrequent; therefore, axillary lymph node resection is not routinely performed unless a palpable axillary lymph node is found or metastasis is confirmed. Metastatic disease develops in 13-40% of patients. When present, spread is hematogenous through stromal cells, affecting the lungs in 66% of cases, the bones in 28% and the liver in 15% of cases after a mean period of 18 months. Most patients with benign or borderline tumors are cured following surgery, while in those in whom the tumor is malignant survival is 94% at 2 years and 87.5% at 5 years [9].

The patient reported here is young and presented with a voluminous and extremely fast-growing breast tumor eight months postpartum, probably as a consequence of hormonal factors. Diagnosis was reached following core biopsy, immunohistochemistry and histopathology of the surgical specimen. These findings reinforce the importance of performing a thorough anamnesis and physical examination for malignancy in cases of more aggressive, larger and fast-growing tumors, even in young patients who are not in the most commonly affected age group. In cases of malignant pathologies, investigation for distant metastasis and patient monitoring for at least three years are imperative due to the high possibility of recurrence.

Conclusion

Phyllodes tumors are rare fibroepithelial tumors of the breast that are classified as benign, borderline or malignant and for which treatment is essentially surgical. Although they are most often benign, histopathological differentiation is important to enable the severity of the case to be established and treatment and follow up to be defined. In cases of malignancy, the possibility of metastases must be thoroughly investigated, since survival rates are high when treatment is appropriate.

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