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# Aggressive malignant phyllodes tumor

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ABSTRACT

*INTRODUCTION:* Originally described in 1838 by Muller, phyllodes tumor is a rare fibroepithelial neoplasm which represents roughly 0.3–0.9% of all breast cancers. Phyllodes tumor are divided into benign, borderline and malignant histologic categories. Malignant phyllodes tumor represent anywhere from 10–30% of all phyllodes tumors. This group has both the potential to recur locally and metastasize, however not all malignant phyllodes behave this way. The challenge lays in predicting which tumor will recur locally or metastasize. Distinguishing this subset of malignant phyllodes tumor is paramount.
*PRESENTATION OF CASE:* We present a case of malignant phyllodes which presented with metastatic disease. What is fascinating about this case is not only the initial presentation but also the aggressiveness of this variation of phyllodes tumor. The patient initially presented with a large mass which encompassed her whole right breast. On surgical pathology the mass measured roughly 31 cm in diameter and weighed over 10 kg. Within 5 weeks from surgery the patient had suffered brain metastases and also 6 local recurrent tumors. The patient passed roughly 11 weeks after her first visit to our office.
*CONCLUSION:* Despite biopsy proven malignant phyllodes tumor, it was near impossible to predict such a rapid course of disease progression in our patient. Our case illustrates the unpredictable nature of this disease in general and it possibly sheds light on a variant of the disease which had undergone an aggressive transformation.



## 1. Introduction

Originally described in 1838 by Muller, phyllodes tumor is a rare fibroepithelial neoplasm. This represents roughly 0.3–0.9% of all breast cancer in women [1,2]. Characterized by a combination of hyper cellular stroma and cleft-like spaces lined by epithelium, phyllodes tumor was classically known as cystosarcoma phyllodes because of the leaf-like projections [3,4]. Renamed phyllodes tumor in the early 1980s, these tumors vary greatly in size and the histological characteristics continue to be the basis for diagnosis and sub classification. Based on established histological criteria PT are classified into three categories – benign, borderline and malignant [5]. The majority of phyllodes tumors are benign and the primary concern after surgical treatment is primarily of local recurrence vs. distant recurrence (Fig. 1a ).

Malignant phyllodes represent anywhere from 10–30% of all phyllodes tumors [6]. There is an abundance of literature regarding invasive ductal carcinoma of the breast and its malignant potential however not as much research has been done on phyllodes tumors. The challenge lays in predicting which tumor will recur locally or metastasize. Phyllodes tumors which fall into histologically benign or borderline category essentially never recur [7]. Phyllodes tumors which fall into a malignant histological category do have a recurrence potential and a metastatic potential on the other hand not all malignant phyllodes tumors recur or metastasize. Distinguishing this subset of malignant phyllodes tumors is paramount. We present a case of malignant phyllodes tumor which presented with metastatic disease, however, what was fascinating about this case was not the as much the initial presentation but the aggressiveness of this variation of phyllodes tumor (Fig. 1b).

## 2. Case report

A 50 year-old woman G3P3 presented with a right breast mass which had been there for 'several years'. A thorough history and physical exam revealed a mass localized to the right breast, which encompassed the entire breast. The mass had increased in size rapidly in the past 6 weeks and it was painful with multiple open wounds. The patient denied other lumps or mass, muscle/bone pain or headaches but complained of fatigue, night sweats and weight change. There was no significant past medical or surgical history or family history (Fig. 1c).

The physical exam, revealed a 'melon-sized' mass throughout entire right breast. There were multiple ulcerations and many engorged veins could be seen throughout breast mass as well. The mass was nodular, hard and was not fixed to the chest wall. There was no lymphadenopathy found (Fig. 2).

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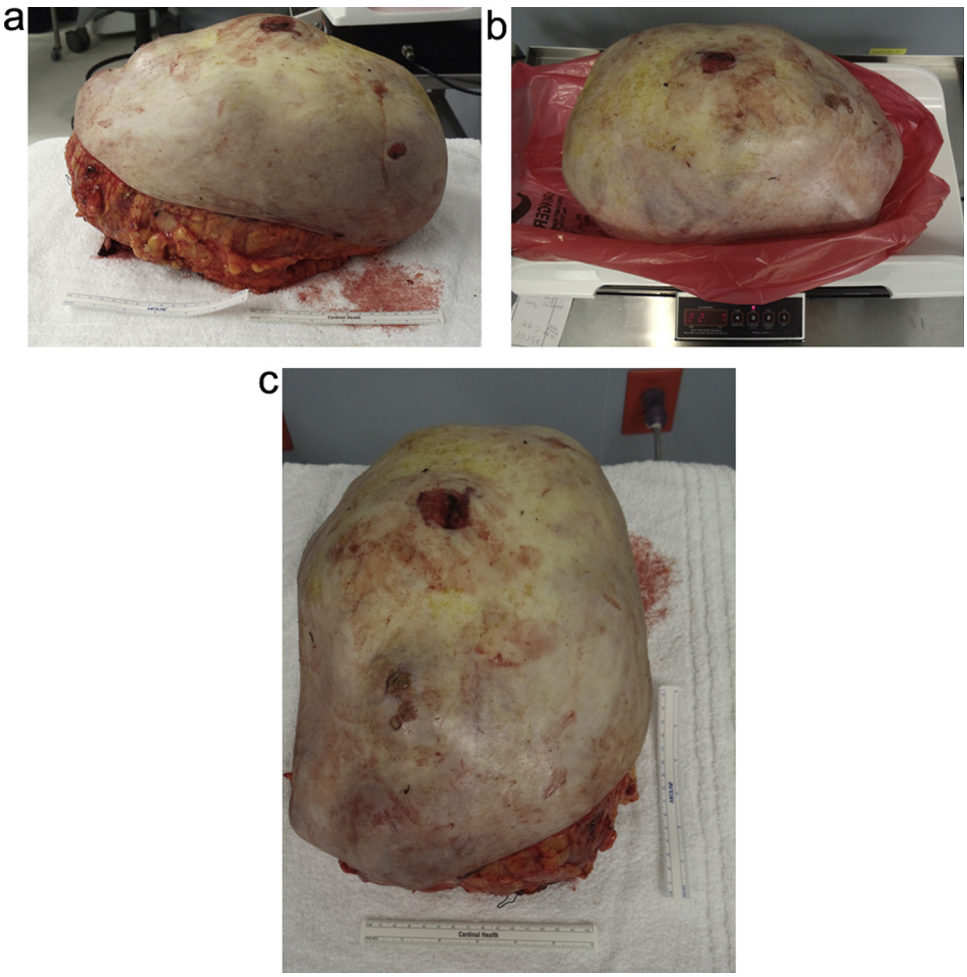


**Fig. 1.** Right mastectomy specimen.

A core needle biopsy was performed in office that day. The biopsy results were found to be a malignant neoplasm with carcinomatous and sarcomatous elements. The cells showed focal marked pleomorphism with >10 mitoses/10HPF. Focal necrosis was seen. The biopsy was estrogen receptor negative and progesterone receptor positive. HER2 was also negative (Fig. 3).

A metastatic workup was performed; this included a CT chest, abdomen, pelvis and also PET scan. The CT of chest was positive for bilateral chest masses which were concerning for metastatic disease as well as a 30 cm mass in the right breast. The patient underwent a right simple mastectomy due to the massive size and ulcerations of the breast. The right breast mass weighed 10.3 kg at the time of excision.

The final pathology from surgery revealed a malignant phyllodes tumor 31.5 cm length × 15.6 cm height. The tumor showed brisk mitotic activity, necrosis, and stromal overgrowth. 1 mm clear margins were attained. A minority of tumor cells in a high grade portion of round cells in the tumor are cytokeratin positive. A small portion had the appearance of myxoid sarcoma. Most of the round cell component is cytokeratin negative; however the positive cells may represent a true differentiation to an epithelial component (carcinosarcoma).

In the immediate post-operative period the patient recovered well. However, within 3 weeks of the mastectomy the patient complained of headaches and was found to have a right parietal mass. The patient underwent a right craniotomy which was positive for a right parietal tumor – metastatic malignant phyllodes tumor. Follow up in our breast clinic one week after was unremarkable and follow up for lung masses was starting. The patient was in clinic roughly 7 weeks from her original presentation with complaints of multiple 'new' lumps on right post-mastectomy site. Physical exam revealed 6 new masses were found, the largest measuring 6 cm and a mass was palpated in the right axilla. A fine needle aspiration was performed. These cells where enlarged pleomorphic, epithelioid cells – identical to original tumor. Before any therapies could be initiated the patient passed shortly after.

## 3. Discussion

As stated prior, one of the challenges facing physicians for patients with phyllodes tumor is predicting which patients will develop local recurrence, metastatic disease or both. The majority of phyllodes tumors present as firm, smooth, well-circumscribed, and rarely painful masses [8]. Most phyllodes tumors occur in women between ages of 35 and 55 years old [9]. Primary treatment for phyllodes tumor is surgical. Depending on the size of the tumor; wide local excision is the treatment of choice. If margins of 1 cm cannot be attained then simple mastectomy is the next best option. Most current studies show that wide local excision with adequate margins yield equivalent results to mastectomy is terms of overall surviva [10,11]. In general, borderline tumors metastasize, however this is not common. In a case review by Moffat et al. only 4% of patients with borderline tumors developed metastatic disease [12]. Malignant tumors develop metastases more commonly. In some studies metastatic disease has been reported between 9 and 32% [13–15]. Limited research and data is available regarding