

Radiagnosis of Primary Breast Sarcoma to Metastatic Malignant Phyllodes Tumor of the Breast

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Abstract:

A tumor with a highly variable course of growth, the phyllodes tumor, can be difficult to differentiate and presents special challenges within staging and grading.¹ Compromising only 0.3-1% of all breast tumors worldwide, phyllodes tumors are fibroepithelial lesions arising from the intralobular stroma of the breast tissue that ranges in their spectrum from benign, borderline, and malignant.¹ Presented here is a rare case of a metastatic malignant phyllodes tumor in a 60-year-old female patient. The lesion was initially identified through self-breast examination and diagnosed as a primary sarcoma through needle core biopsy. Following the removal of the tumor by radical mastectomy, further histologic review rediagnosed this tumor as a malignant phyllodes tumor due to the presence of an epithelial component not previously identified. Accurate diagnosis of a phyllodes tumor is pivotal given the high rate of recurrence and metastasis in malignant tumors. The varying morphology and subjectivity to grading of phyllodes tumors requires extensive sectioning by the pathologists' assistant for the proper diagnosis and subsequent treatment to be rendered.

Background:

A 60-year-old Caucasian woman palpated a mass in her lateral left breast which grew rapidly until she sought medical care when pain started 4 months later. Needle core biopsy revealed a high grade sarcomatoid lesion (**Figure 1**) with a differential diagnosis of liposarcoma, metastatic tumor, or angiosarcoma. The decision was made for surgical excision by radical mastectomy. The presence of an epithelial component was noted after surgical excision, changing the diagnosis from a primary sarcoma to a malignant phyllodes tumor. Wide tumor free surgical margins were present at time of gross and histologic evaluation. In the 21 months following surgical excision the patient presented with recurrence on the left chest wall with metastasis to the lungs, pancreas, liver, and brain.

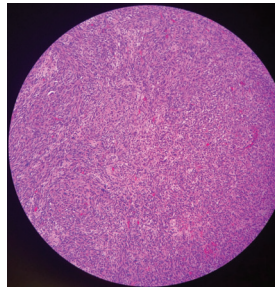


Figure 1: Photomicrograph from needle core biopsy of left breast mass demonstrating spindle cell morphology.



Figure 2: Gross photograph of the left breast following mastectomy showing ulceration through the anterior skin surface.

Gross Dictation:

SPECIMEN A:

LABELED-FIXATIVE: "Left breast with skin, short stitch superior, long stitch lateral, in formalin per surgeon"/formalin (cold ischemia time: 58 minutes; fixation began at 1558, total fixation is 23.5 hours).

TISSUE DESCRIPTION: 827.40 g mastectomy, posterior/deep margin demonstrates smooth fascial plane with focal areas of muscle; specimen inked as follows: superior-anterior aspect blue, inferior-anterior green, and posterior/deep margin black.

SIZE:

Superior to inferior: 16.0 cm,
Medial to lateral: 18.0 cm,
Anterior to posterior: 5.2 cm.

SKIN: 18.0 x 16.2 cm portion of tan skin and nipple. 4.0 x 4.0 cm area of discoloration and nodularity appears 3.8 cm lateral to nipple, with a 1.0 x 0.7 cm central area of ulceration. The nodular ulcerated area comes within 8.2 cm from the superior skin margin, 5.0 cm from the inferior skin margin, 6.2 cm from the lateral skin margin and 10.0 cm from the medial skin margin. (**Figure 2**)

TUMOR DESCRIPTION: Pink-white whorled mass with multiple focal areas of hemorrhage and central necrosis (70%). Mass grossly involves the anterior skin, communicating with the area of nodularity and ulceration. Mass measures 10.0 cm from superior to inferior, 8.5 cm from medial to lateral, and 5.2 cm from anterior to posterior. Mass abuts but does not grossly involve muscle on posterior aspect. Biopsy site identified within mass. (**Figure 3**)

LOCATION: Central portion of breast.

PROXIMITY TO MARGINS:

Posterior/deep soft tissue margin: 0.2 cm.
Anterior superior soft tissue margin: 2.0 cm.
Anterior inferior soft tissue margin: 3.5 cm.
Lateral soft tissue margin: 5.5 cm.
Medial soft tissue margin: 7.5 cm.
Superior skin margin: 2.0 cm.
Inferior skin margin: 3.5 cm.
Lateral skin margin: 4.7 cm.
Medial skin margin: 6.2 cm.

REMAINING CUT SURFACE: Yellow-tan lobulated fibroadipose tissue.

INTRAMAMMARY LYMPH NODES: None grossly identified.

Representative sections are submitted as follows:

A1: Perpendicular section of nipple.

A2: Mass involving anterior skin.

A3-A4: Mass in relation to posterior margin.

A5: Perpendicular section of anterior superior soft tissue margin/superior skin margin.

A6: Perpendicular section of anterior inferior soft tissue margin/inferior skin margin.

A7: Perpendicular section of lateral skin margin/lateral soft tissue margin.

A8: Perpendicular section of medial skin margin/medial soft tissue margin.

A9-A18: Representative sections of mass.

A19-A20: Central necrosis within mass.

SPECIMEN B:

LABELED-FIXATIVE: "Left axillary lymph node", formalin.

TISSUE DESCRIPTION: 4.0 x 3.0 x 1.2 cm fragment of yellow-tan adipose tissue, three possible lymph nodes identified.

LYMPH NODE SIZE: 0.5-2.2 cm in greatest dimension.

All lymph nodes are submitted as follows:

B1: One lymph node, bisected.

B2: One lymph node.

B3-B4: One lymph node, bisected.

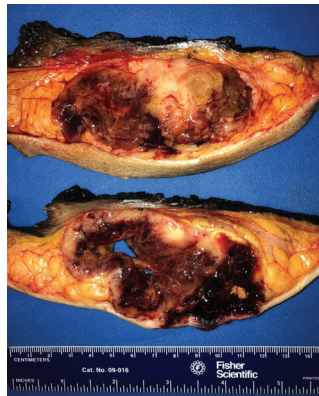


Figure 3: Cut surface of the left breast showing necrosis involving most of the tumor.

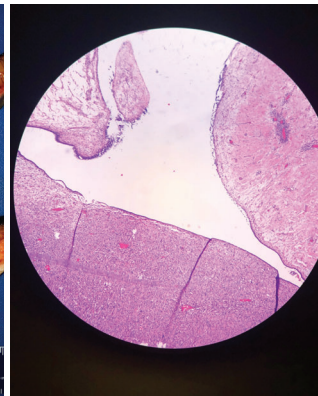


Figure 4: Representative photomicrograph from the left breast mass post-mastectomy demonstrating the epithelial component required for a phyllodes tumor diagnosis.

Final Diagnosis:

Histologic assessment of hematoxylin and eosin (H&E) slides revealed high grade pleomorphic sarcoma set on a background of phyllodes tumor. Some tumor sections showed residual leaf-like structures lined by benign epithelium and overgrown by hypercellular stroma (**Figure 4**). Most sections demonstrated high grade pleomorphic sarcoma with necrosis, marked nuclear pleomorphism and brisk mitotic activity. It is noted that unlike the needle core biopsy the mass clearly contains benign epithelial elements, rendering the diagnosis of this tumor a malignant phyllodes tumor with ulceration of skin. All margins are free of malignancy.

Conclusion:

Phyllodes tumors of the breast are rare fibroepithelial neoplasms which recur frequently yet have a low rate of metastasis.² In the United States, these tumors account for approximately 0.3-0.5% of all breast cancers.^{3,4} As evident in this case, phyllodes tumors typically arise in women in their 6th decade of life and present as a lump found during a self-breast exam.⁵ They have a biphasic growth pattern with a long initial slow growing phase before a sudden rapid increase in size over weeks or months, causing its clinical presentation.^{3,4} Due to a lack of yearly breast imaging, it is not possible to confirm whether the phyllodes tumor in our presenting case developed from a previous tumor or if it was a new neoplasm. However, over 4 months the lesion went from a palpable lump to a 10.0 cm mass, consistent with rapid biphasic growth.

Though rapid growing, 90% of phyllodes tumors are classified under benign or borderline, with the remaining 10% of malignant cases metastasizing 22% of the time, typically to the lungs and bones and rarely to other locations.⁴ Recurrence rates are significantly higher for tumors larger than 10 centimeters, reaching 41% and necessitating aggressive treatments and wide resection margins.⁶ Excisional surgery is currently the gold standard treatment of all grades of phyllodes tumors, with large tumors requiring a complete mastectomy including margins of 1 cm or greater.^{5,7} As a result of the high risk of recurrence with malignant phyllodes tumors, their excision is typically more extensive though it rarely involves an axillary tail dissection due to the low rate of lymphatic spread.⁵ Despite the pre surgical screening for metastasis and wide, tumor-free margins, this case had rare spread to the lungs, liver, pancreas, chest wall, and brain after surgical intervention. It is feasible that there was a recurrence of the original phyllodes tumor appearing as the lump on the patient's chest wall 11 months after her mastectomy, which subsequently spread hematogenously and lead to her ultimate demise.

Evaluation of phyllodes tumors typically begins with needle core biopsy, though results are often inconclusive or incorrectly diagnosed due to the benign and malignant phyllodes tumor close relation to fibroadenomas and sarcomatous lesions, respectively.⁷ Undifferentiated mammary sarcomas are morphologically indistinguishable from malignant phyllodes tumors on core biopsy, particularly when no epithelial component is present, such as in this case.⁵ The differential for a malignant phyllodes tumor will most commonly involve metastatic carcinoma and primary sarcoma of the breast.⁸

When grossing tumors with a differential including phyllodes tumor, the pathologists' assistant should take extra caution in sampling the tumor extensively to ensure at least one slide contains the epithelial component for proper differentiation. These grossing guidelines are of extra importance for soft tissue tumors diagnosed by needle core biopsy, as staging and treatment options differ between various sarcomatous lesions.¹ Histologically, the malignancy of a phyllodes tumor is based on the appearance of nuclear pleomorphism, abundant mitosis, and stromal overgrowth in the presence of a leaflike structure. Distant metastasis generates a poor prognosis due to the lack of response to chemotherapy, with most patients passing within 3 years of initial treatment.² Early detection and surgical intervention are currently the best treatment for malignant phyllodes tumors. Further research is needed to investigate the pathogenesis and risk factors associated with phyllodes tumors.

References:

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