

RADIOLOGIC-PATHOLOGIC CORRELATION

Malignant Phyllodes Tumor with Chondrosarcomatous Differentiation: Radiological-Pathological Correlation

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ABSTRACT

We present a case of a 63-year-old woman with malignant phyllodes tumor in her left breast. On imaging, a large, dumbbell-shaped, predominantly cystic mass with thin peripheral enhancement was noted. The lesion was causing rib destruction, chest wall invasion, and intrathoracic extension. These aggressive imaging features were considered highly suspicious of a malignant chest wall tumor. Subsequent chest wall resection of the tumor showed breast tissue with a biphasic lesion composed of proliferated spindle cells in loose sheets with extensive islands of atypical cartilage and a scanty epithelial component, including compressed ducts in the periphery of the lesion. A diagnosis of a malignant phyllodes tumor with stromal overgrowth and chondrosarcomatous differentiation was made in view of the presence of a benign epithelial component and negative reaction of the stromal component with a pancytokeratin. To the best of our knowledge, a phyllodes tumor with the radiological features of chest wall invasion and intrathoracic extension has not been described in the literature until now. Malignant phyllodes should be included in the list of differentials along with sarcomas on encountering lesions with such aggressive imaging features.

Key words: Chondrosarcomatous differentiation, chest wall erosion, intrathoracic extension, malignant phyllodes, phyllodes tumor

INTRODUCTION

Phyllodes tumors are rare entities and constitute only 0.3-1.0% of all breast tumors.^[1,2]

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Also known as cystosarcoma phyllodes, they are biphasic proliferations with an epithelial component composed of broad leaf-like papillae in a cellular stroma. They are graded as benign, borderline, and malignant based on stromal characteristics.^[3] A 63-year-old female presented with a large, firm to hard left breast mass that had been present for 1 year. The lesion had been smaller earlier and had grown rapidly over the last few weeks. On examination, the skin over the mass appeared tense with prominent veins. No evident ulceration was seen. After clinical examination, a provisional diagnosis of chest wall sarcoma was made.

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RADIOLOGIC FEATURES

Ultrasound (US) revealed a thin-walled, predominantly cystic mass with dense internal echoes and echogenic debris within [Figure 1].

Patient underwent a contrast-enhanced computed tomography (CECT) scan on a 16-slice CT scanner. CECT was performed by injecting 80 ml of non-ionic intravenous contrast Iohexol through an 18-gauge needle in the antecubital vein at a rate of 3 ml/s. Acquisition was done at a slice thickness of 2.5 mm. In addition to a baseline non-enhanced scan, image acquisition was done during venous phase (70 s). The 2.5-mm-thick axial images were reformatted into thinner sections in three orthogonal planes. CT revealed a large, dumbbell-shaped, lobulated mass lesion in the left chest with a large extrathoracic (measuring 13.3 × 7.6 × 8.3 cm) and an intrathoracic component (measuring 4.6 × 5.7 × 9.4 cm). The lesion showed thin peripheral enhancement with extensive central necrosis [Figures 2 and 3]. Multiple specks of calcification were also noted within the lesion, along with few enhancing incomplete septae. The mass eroded the anterior aspects of left 3rd, 4th, and 5th ribs as well as the pectoral muscle. The intrathoracic component was seen to compress the mediastinum, causing a mild contralateral shift. However, a clear fat plane was seen separating the lesion from the mediastinal structures. The underlying lung parenchyma was also intact.

PATHOLOGIC FEATURES

The tumor was resected along with the three involved ribs. A large, dumbbell-shaped lesion was noted, extending

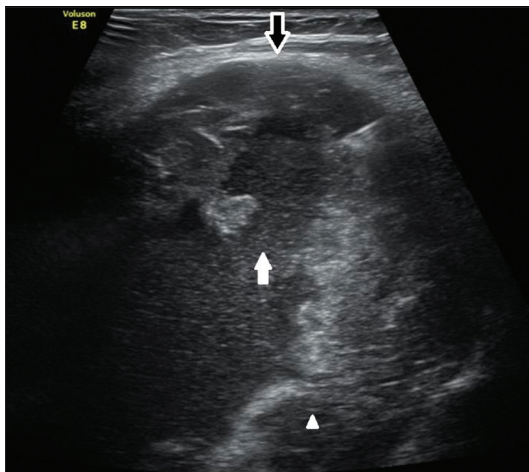


Figure 1: 63-year-old female presented with a large mass in the left breast and was diagnosed with malignant phyllodes with chondrosarcomatous differentiation. B-mode ultrasound image demonstrates predominant cystic nature of the lesion (black and white arrow) with internal echoes (solid white arrow) and debris (white arrowhead).

anterior and posterior to the ribs [Figure 4]. The tumor measured 16 × 13 × 9 cm and appeared fairly well circumscribed. On sectioning, the portion of lesion anterior to ribs showed extensive cystic degeneration, while the posterior part was firm with gelatinous foci and areas of calcification (which corroborated with the imaging appearance).

Histologic examination of the tissue with hematoxylin and eosin (H and E) stain revealed breast tissue with a biphasic lesion composed of proliferated spindle cells in loose sheets with extensive islands of atypical cartilage and a scanty epithelial component, including compressed ducts in the periphery of the lesion [Figure 5]. The stromal component exhibited moderate cytological atypia with increased mitosis and giant tumor cells [Figure 6]. The cartilaginous component included atypical and binucleated chondrocytes [Figure 7]. The final diagnosis was a malignant phyllodes tumor with stromal overgrowth and chondrosarcomatous differentiation.

The patient underwent chest wall resection followed by radiotherapy. Three-month follow-up CECT post surgical resection and radiotherapy showed no evident residual or recurrent lesion at the surgical site [Figure 8].

DISCUSSION

First described by Johannes Müller in 1838 as cystosarcoma phyllodes, phyllodes tumor is a rare fibroepithelial neoplasm. It is graded as benign, borderline, and malignant based on stromal characteristics.^[3] Approximately 5–25% of phyllodes tumors are found to be malignant.^[4]

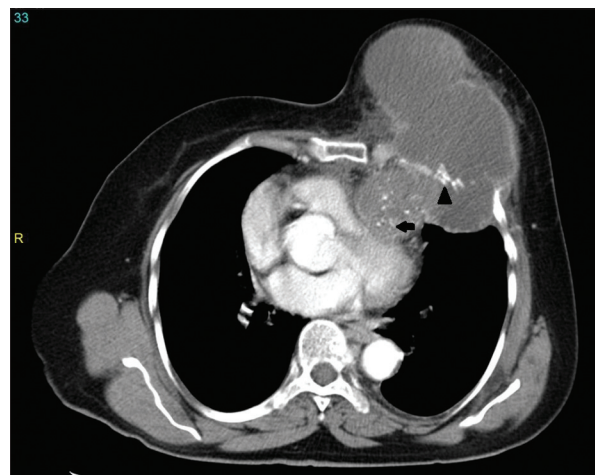


Figure 2: 63-year-old female presented with a large mass in the left breast and was diagnosed with malignant phyllodes with chondrosarcomatous differentiation. Axial contrast-enhanced CT image reveals the predominantly cystic nature of the tumor with rib destruction (black arrowhead), intrathoracic extension and compression of the underlying mediastinal structures without invading them. Calcific specks are noted in the intrathoracic component (black arrow).

On US, phyllodes tumor usually appears as a well-defined mass with heterogeneous internal echoes sometimes having posterior acoustic enhancement.^[5] Liberman et al.,^[6] reported that a phyllodes tumor with diameter greater than 3 cm tends to be associated with malignancy. However, there are no reliable mammographic or US features to differentiate benign from malignant phyllodes tumor.^[7] On CT, a phyllodes tumor can appear as a heterogeneous enhancing mass which may contain solid and cystic components including irregular enhancing septations.^[8] However, findings on imaging studies are not considered pathognomonic of phyllodes tumors. Chest wall invasion and intrathoracic extension have not been described in the context of phyllodes until now.

In our case, important imaging differentials included chest wall sarcoma, chest wall metastasis, and breast

carcinoma (infiltrating ductal variety). Malignant chest wall tumors usually arise from the chest wall musculature, ribs, or pleura. They typically manifest as large, poorly marginated, infiltrative masses and are usually symptomatic.^[9] Chondrosarcoma is the most common malignant primary tumor of the chest wall. The characteristic CT appearance of chondrosarcoma consists of a well-defined, lobulated soft-tissue mass with foci of chondroid matrix calcification.^[10] Different patterns of chondroid calcifications described are “popcorn” type, ring and arc, and stippled variety. Bone destruction and invasion of overlying soft tissue may also exist. In our case, considering the large size of tumor, rib destruction, stippled areas of calcification, and heterogeneous enhancement, chest wall chondrosarcoma possibly arising from the ribs was kept as the first differential



Figure 3: 63-year-old female presented with a large mass in the left breast and was diagnosed with malignant phyllodes with chondrosarcomatous differentiation. Sagittal contrast-enhanced CT chest demonstrates the intrathoracic component of the tumor (white arrow). Stippled pattern of calcification is noted in the intrathoracic component (black arrows).

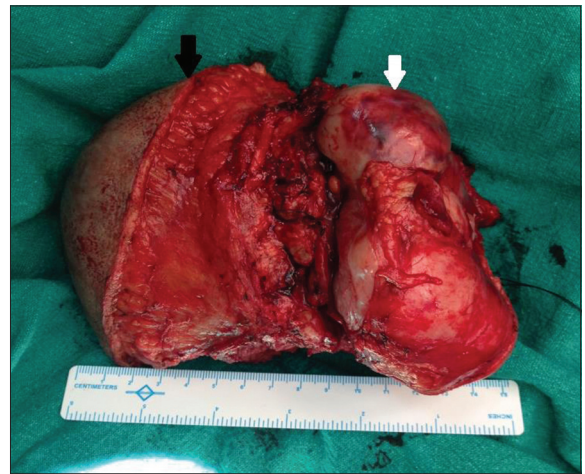


Figure 4: 63-year-old female presented with a large mass in the left breast and was diagnosed with malignant phyllodes with chondrosarcomatous differentiation and underwent chest wall resection. Postoperative photograph of the resected specimen demonstrates the extrathoracic (black arrow) and intrathoracic (white arrow) parts of the tumor.

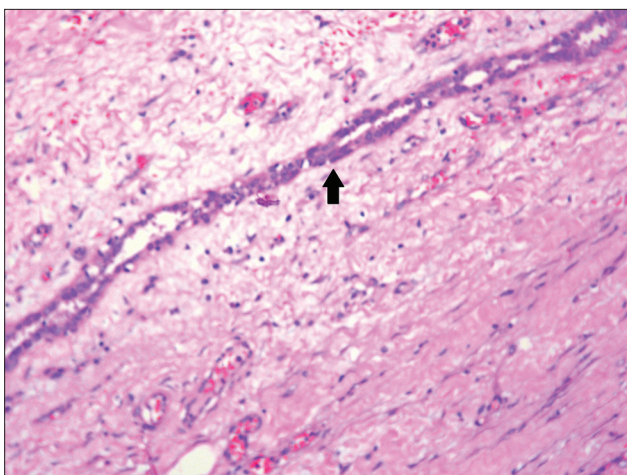


Figure 5: 63-year-old female presented with a large mass in the left breast and was diagnosed with malignant phyllodes with chondrosarcomatous differentiation. Hematoxylin and eosin stained tissue (×10) demonstrates epithelial component with duct lined by bilayered epithelium (arrow).

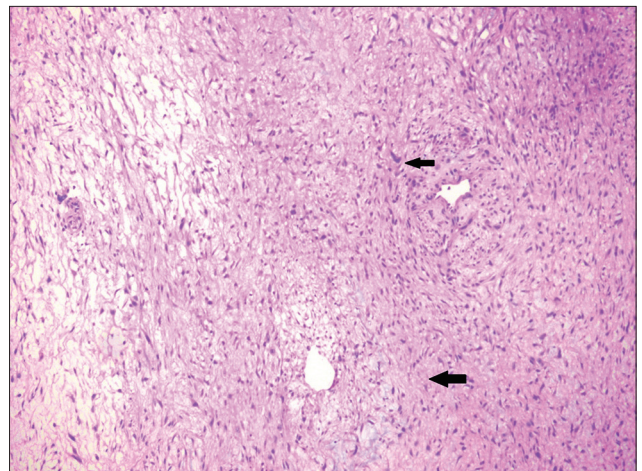


Figure 6: 63-year-old female presented with a large mass in the left breast and was diagnosed with malignant phyllodes with chondrosarcomatous differentiation. Hematoxylin and eosin stained tissue (×10) demonstrates stromal component including spindle cells (arrows) exhibiting moderate cytological atypia and increased mitoses.

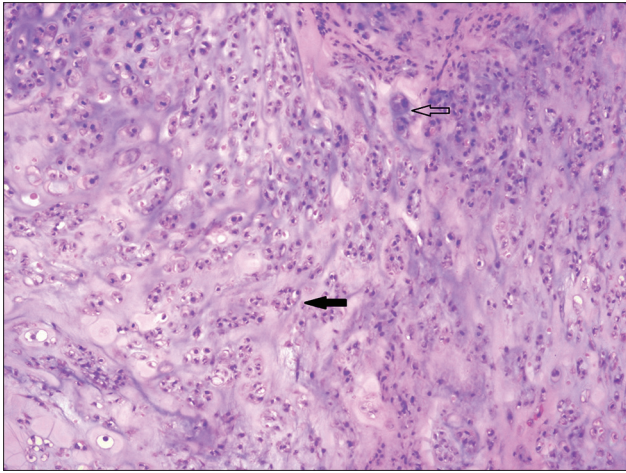


Figure 7: 63-year-old female presented with a large mass in the left breast and was diagnosed with malignant phyllodes with chondrosarcomatous differentiation. Hematoxylin and eosin stained tissue ($\times 10$) reveals chondrosarcomatous component with cartilage, including binucleate and atypical chondrocytes with hyperchromatic nuclei (arrows).

diagnosis. Chest wall metastases can have similar imaging appearance. However, in the absence of a known primary, metastasis was considered less likely. Breast carcinomas (infiltrating ductal variety), on the other hand, are clinically more likely to cause fungation and ulceration. They appear more solid, have spiculated margins, and are usually associated with axillary lymphadenopathy.^[8]

A diagnosis of a malignant phyllodes tumor with stromal overgrowth and chondrosarcomatous differentiation was made on histopathological examination. The presence of a benign epithelial component and negativity of the stromal component with a pancytokeratin excluded the possibility of breast carcinoma and chondrosarcoma.

Treatment of phyllodes tumor requires complete surgical excision with wide margins.

CONCLUSION

Malignant phyllodes tumor can rarely be associated with a chondrosarcomatous differentiation, and thus mimics a chest wall tumor clinically and radiologically. Malignant phyllodes should be included in the list of differentials along with sarcomas on encountering lesions with chest wall invasion and intrathoracic extension on imaging. Final diagnosis of this rare entity can be made only on excision biopsy.

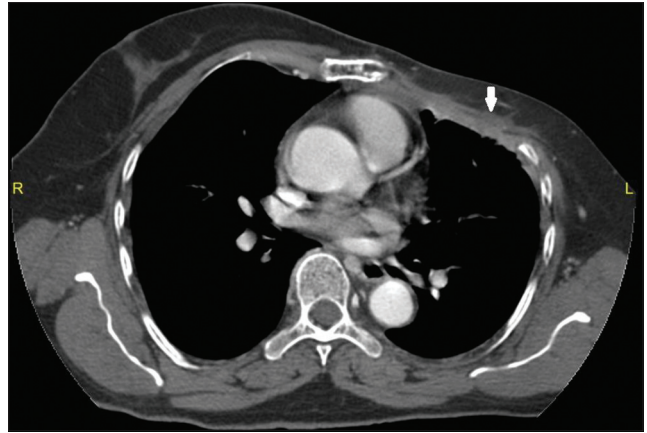


Figure 8: 63-year-old female presented with a large mass in the left breast and was diagnosed with malignant phyllodes with chondrosarcomatous differentiation. Three-month follow-up contrast-enhanced CT post surgical resection and radiotherapy shows no evident residual or recurrent lesion at the surgical site (white arrow).

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