

Case Report

Recurrent phyllodes tumor of the breast transforming to a fibrosarcoma

Gopalakrishnan Gunasekaran^{1*}, Debasis Naik¹, Sonam Sharma², Vimal Bhandari¹,
Ashish Kumar Mandal², Deepak Rajput³

¹Department of General Surgery, Vardhman Mahavir Medical College and Safdarjung Hospital, New Delhi, India

²Department of Pathology, Vardhman Mahavir Medical College and Safdarjung Hospital, New Delhi, India

³Department of Surgery, Vardhman Mahavir Medical College and Safdarjung Hospital, New Delhi, India

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*Correspondence:

Gopalakrishnan Gunasekaran,

E-mail: gpgopslakrishnan1987@gmail.com

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ABSTRACT

Phyllodes tumors are fibroepithelial tumors of the breast, representing 2-3% of all fibroepithelial tumors, and <1% of all breast tumors. Malignant transformation may occur usually within stromal component of phyllodes tumor. Careful characterization of the stromal compartment is critical, since it is the pathologic features of stromal cells that determine its malignant potential. Primary sarcomas of the breast are extremely rare and account for <0.1% of all malignant tumor of the breast, of which the conversion of a phyllodes tumor to sarcoma of the breast is even very rare and only two cases have ever been reported. We present a case of recurrent phyllodes tumor of the breast transforming to a fibrosarcoma.

Keywords: Fibrosarcoma, Phyllodes tumor, Breast

INTRODUCTION

Phyllodes tumors are rare fibro-epithelial lesions that account for under 1% of all breast neoplasms.^{1,2} Based on histological characteristics of tumor, including its surgical margin, stromal cellularity, stromal overgrowth, tumor necrosis, cellular atypia and the number of mitoses per high power field (HPF), it is classified into benign, borderline and malignant categories.³ Primary sarcomas of the breast are extremely rare and account for <0.1% of all malignant tumors of the breast, of which the conversion of a phyllodes tumor to sarcoma of the breast is even very rare and only two cases have ever been reported.⁴

CASE REPORT

A 40-year-old lady presented to our outpatient department with a rapidly growing, painless lump in the upper and inner quadrant of right breast for 6 months. There was no associated nipple discharge. She had a history of lumpectomy 2 years back for a similar lump in the same

region of the breast. On examination, 4 cm × 4 cm lump was noted in the upper and inner quadrant of the right breast, which was fixed to the skin at the site of previous scar and free from pectoralis muscle and chest wall. Fine needle aspiration cytology revealed a phyllodes tumor. Wide local excision was done.

Gross specimen showed a firm, well-circumscribed, round mass that was gray-white with areas of hemorrhage (Figure 1). Histopathological examination of which showed stromal overgrowth and hypercellularity, nuclear atypia and increased mitotic count (6-8/HPF). The cells were spindle-shaped and varied little in size and shape, had scanty cytoplasm with indistinct cell borders and were separated by interwoven collagen fibres arranged in a parallel fashion, favouring the diagnosis of fibrosarcoma (Figure 2). There were also focal areas of borderline phyllodes tumor. Immunohistochemistry showed that the tumor cells were positive for vimentin and focal positive for smooth muscle actin (Figure 3). After observing these histopathological features of the tumor, her previous slides

were reviewed. Histopathological examination of which showed a tumor with the stromal hypercellularity and the presence of benign glandular elements. Although the stroma was more cellular, the tumor cells showed mild pleomorphism and mitoses were infrequent. These features were suggestive of a borderline phyllodes tumor (Figure 4). All the resected margins were free of the tumor. We finally came to the conclusion that phyllodes tumor of the breast had transformed to fibrosarcoma. Patient was then given radiotherapy followed by five cycles of chemotherapy consisting of cyclophosphamide, vincristine and adriamycin. Patient was followed for 1 year with no recurrence.

DISCUSSION

‘The phyllodes tumor, firstly described by Johannes Muller in 1838 has presented a diagnostic and treatment dilemma for physicians since its original description. Classically, the name cystosarcoma phyllodes was assigned because of the tumor’s fleshy appearance and tendency to contain macroscopic cyst. The term however is a misnomer as

these tumors are usually benign. Phyllodes tumor is the currently accepted nomenclature according to the World Health Organization.’⁵

Phyllodes tumors are fibroepithelial tumors of the breast, representing 2-3% of all fibroepithelial tumors, and <1% of all breast tumors.⁶ Phyllodes tumors are uniquely found in breast tissue and histologically have both an epithelial component as well as metaplastic mesenchymal (stromal) component. Clonal analyses have revealed that some phyllodes tumor develop from fibroadenomas.⁷

Phyllodes tumor, despite being benign, has a tendency to recur after surgery. An originally histologically benign tumor may develop malignant features with recurrence.⁸ The literature shows that in most cases more aggressive growth and enhanced malignancy is found on recurrence.⁴ Recurrence may result from proliferative remnants of the primary tumor following local excision or they may

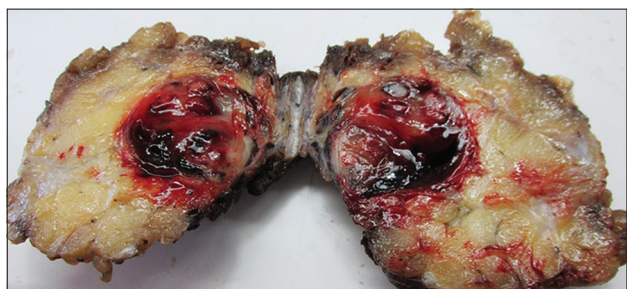


Figure 1: Gross specimen showing a firm, well-circumscribed, round mass that is gray-white with areas of hemorrhage.

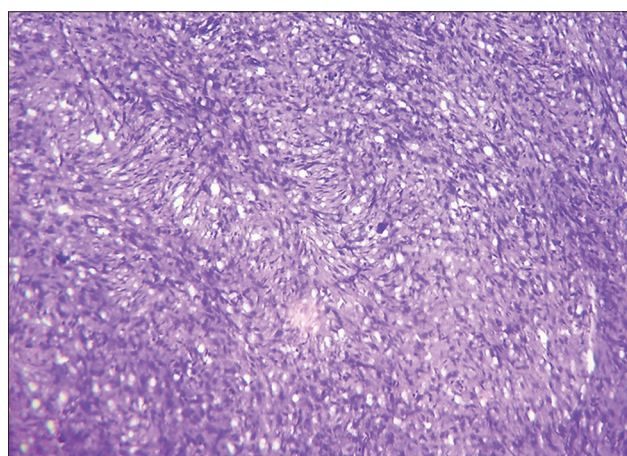


Figure 2: Slide showing stromal overgrowth and hypercellularity, nuclear atypia and increased mitotic count (6-8/high power field). The cells are spindle-shaped and vary little in size and shape, have scanty cytoplasm with indistinct cell borders and are separated by interwoven collagen fibers arranged in a parallel fashion, favoring the diagnosis of fibrosarcoma.

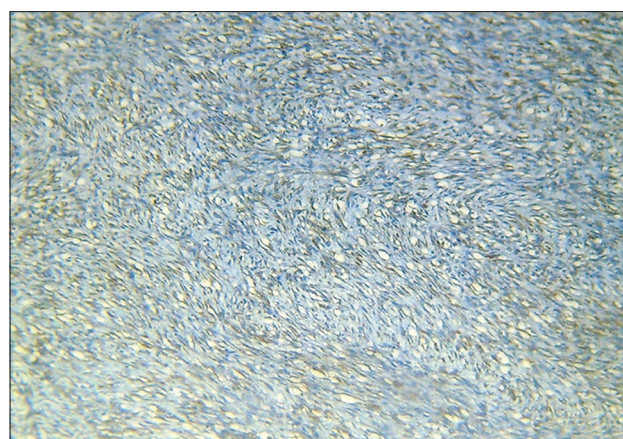


Figure 3: Immunohistochemistry study showing, the tumor cells positive for vimentin.

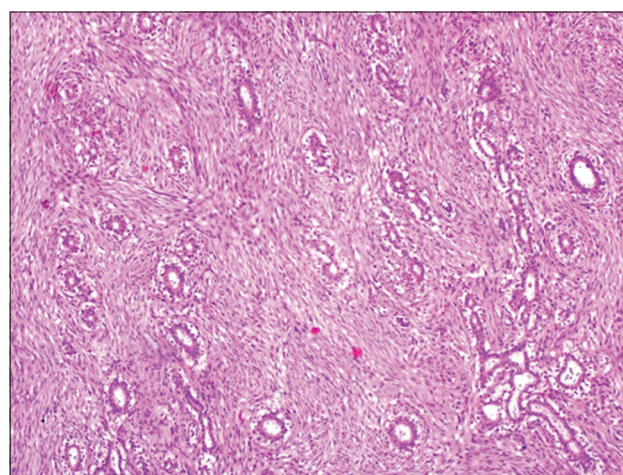


Figure 4: Microphotograph of first surgical specimen slides showing moderate stromal hypercellularity with mild pleomorphism of the spindle cells and stromal overgrowth with mild atypical epithelial hyperplasia, suggestive of phyllodes tumor (borderline type) (H & E, ×10).

be *de-novo* tumors induced by an extra-tumoral stromal hypercellularity resulting in new benign cystosarcoma phyllodes.⁹

“It is also well known that phyllodes tumors may display increased cellularity with pleomorphism, nuclear atypia and malignant transformation of stroma, rendering them borderline or malignant phyllodes tumors. The most common patterns of malignant transformation are fibrosarcoma and myxoliposarcoma. Other differentiation patterns include chondrosarcoma, osteosarcoma and leiomyosarcoma. Frequently, areas of malignant transformation are focal and may be overlooked if careful sampling is not performed.”¹⁰

Patients typically present with a single, round or ovoid-shaped, painless lump. Generally, it is smooth, mobile, well circumscribed and not involving the nipple. It is more than 5 cm in diameter, located in the center of the breast except giant lumps that can occupy the entire breast. The lump exhibits continuous growth, but it may also increase rapidly in size. As a result of stretching and increased pressure, the attenuated skin ulcerates secondarily to ischemia.¹¹ The most common site of the initially diagnosed distant metastasis was the lung. Other metastatic sites include the bones, liver, heart, and distant lymph nodes.¹² Rarely, direct extension without distant metastasis may result in death.^{13,14} As with soft tissue sarcomas, in general, distant pulmonary metastasis may be resectable for a possible cure if it is the only site of distant disease.^{15,16}

“Elson et al. reported the mammographic findings of five cases with fibrosarcoma of the breast. In their study, the mammograms showed high-density masses with margins varying from poorly defined to well-defined and the diameter ranged from 1.5 cm to 7.0 cm. Calcified osseous elements were present in one of the masses. However, these findings were nonspecific.”¹⁷

Ultrasonography shows the tumor to be nodular, well-circumscribed, hypoechoic, heterogeneous lesion, and blood flow may be found. Cystic echo may be seen in tumor. Most of cystic echo is due to hemorrhage, necrosis and mucoid degeneration. Computed tomography (CT) scan has been widely used in recent years in the diagnosis of soft tissue sarcoma, providing tumor size, location and anatomical relationship with adjacent tissues and organs. Magnetic resonance imaging (MRI) has also been used. Comparing with CT, MRI has more significance in detecting the relationship between tumors and surrounding tissue.¹¹

Histologically, tumor is very cellular, exhibiting sheets and fascicles of spindle cells, at places forming storiform and herringbone pattern with nuclear pleomorphism, atypia and increased mitoses.¹⁸ Van Geison stain shows large areas of collagen fibers arranged in intertwining whorled bundles and that also surround individual cells with tumor necrosis. On immunohistochemistry, the tumor cells are positive for vimentin I collagen and negative for S-100 protein, epithelial membrane antigen, keratin and binding protein.¹¹

Studies suggest that if negative margins can be achieved, then breast sarcomas should be managed by conservative surgery with post-operative irradiation with dose of 50 gray to the whole breast, and at least 60 gray to the tumor bed.⁴ Tumor size may be more important prognostic factor than tumor grade. As large tumor size and positive surgical margins incur a higher risk for local failure, radiotherapy is probably indicated in these selected cases and should be given within 4 months of surgery. Radiation treatment alone after mastectomy may fail to prevent a recurrence from the original tumor, whether benign or malignant. Chemo-radiation may have a role in large tumors when an aggressive approach is required as in any soft tissue sarcoma whereas surgery with or without radiotherapy remains the standard treatment.⁴

CONCLUSION

Fibrosarcomatous transformation of recurrent phyllodes tumor of the breast is rare. Till now only two cases have been reported in the literature. Because of this, only limited data regarding presentation, diagnosis and treatment of this tumor is available. If a tumor of similar aspect occurs in the breast following excision of phyllodes tumor, one should consider the possibility of sarcomatous transformation and careful approach is needed in early.

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