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Case Report

MALIGNANT PHYLLODES WITH OSTEOSARCOMATOUS DIFFERENTIATION – A CASE REPORT

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Abstract

Introduction: Malignant phyllodes tumor is a rare and aggressive neoplasm, which presents as breast lump and is detected on routine examination in premenopausal women. Heterologous differentiation is uncommon in these tumors. Owing to its rare occurrence, very few treatment protocols have been formulated for these cases posing a surgical dilemma.

Presentation of Case: For a better understanding of the pathogenesis and prognosis of these tumors we discuss here the clinical profile of a 49 year-old lady with metastatic phyllodes tumor of the breast with osteosarcomatous differentiation.

Discussion: Malignant phyllodes with osteosarcomatous differentiation are highly aggressive and extremely rare constituting 1.3% of all breast cancers. It has been hypothesized that malignant change of metaplastic stromal result in these neoplasms.

Conclusion: To conclude, malignant phyllodes are a rare entity. Osteosarcomatous differentiation is almost unheard of and indicates an aggressive biological behavior. Accurate diagnosis and timely surgical management is essential to prevent progression of the disease and improve survival.

Key Words: Breast tumor, malignant phyllodes tumor, osteosarcomatous features, infiltrating ductal carcinoma.

Introduction:

Phyllodes are a rare stromal epithelial tumor of the breast accounting for less than 1 percent of breast tumors, over half of which are benign. Based on mitotic index, phyllodes are classified as benign, borderline and malignant. Malignant phyllodes may contain heterologous stromal components such as liposarcoma, rhabdomyosarcoma, chondrosarcoma or osteosarcoma. [1-2]

Osteosarcomatous differentiation is rare and very little is known about its molecular profile. [3-5]

Malignant phyllodes are best managed with wide local excision. Very large malignant phyllodes may require mastectomy. In most cases, radiation therapy is not required.

Axillary dissection is not routinely recommended, as metastasis is rare.

We report a case of malignant phyllodes with osteosarcomatous differentiation with metastasis to the contralateral breast, in a premenopausal lady.

Case Report:

A 49 year-old lady was referred to our hospital, post lumpectomy, for a non-tender lump in the left breast. Clinical examination, on the first visit, showed a healed scar with no residual, palpable lump in the upper outer quadrant of the left breast. There was no significant axillary lymphadenopathy. The right breast and axilla were normal clinically. Mammography showed postoperative changes in the left breast with no residual lump in the left and a normal appearing right breast. Histopathological review of the sections revealed a phyllodes tumor (Figure 1) with cystic changes. The resected margins were negative. She was kept on follow-up and explained about self-breast examination (SBE) and to report if any new palpable lumps or fresh symptoms developed. Within eight months, she presented with a painless lump in the left breast (previous operative scar). Examination revealed a 3 x 2 cm hard lump in the upper outer quadrant of the left breast, just below the previous operative scar, with minimal mobility. Clinically, no significant lymphadenopathy was noted. In view of her previous history, a recurrence of the primary disease was suspected. Mammography showed a BIRADS IV lesion. Fine needle aspiration cytology rendered a diagnosis of phyllodes tumor and a wide local excision of the lesion was performed. Histopathologically, a diagnosis of malignant phyllodes with osteosarcomatous differentiation over a background of infiltrating ductal carcinoma (IDC) was rendered with negative margins. (Figure 2)

A completion mastectomy was offered, since very minimal breast tissue was left. In view of microscopic evidence of ICD, axillary clearance was also done in a second sitting. No residual tumor or lymph node metastasis was identified.

Two months post mastectomy, she developed a lump in the right breast. On physical examination, a hard, mobile lump measuring 4 x 5 cm was detected located in the upper outer quadrant and measured 4 x 5 cm, with no evidence of skin infiltration. Multiple enlarged right axillary lymphnodes were also palpable. Sono-mammography demonstrated a BIRADS IV lesion. Fine needle aspiration cytology of the lesion demonstrated a high grade infiltrating ductal carcinoma (IDC) with dense lymphocytic infiltration. Magnetic resonance imaging (MRI) chest was done to look for recurrent disease in the left remnant breast tissue and to rule out multicentric disease in the right breast. Patient underwent right breast conservation surgery. Final histopathology report revealed metastatic malignant phyllodes with osteo-sarcomatous differentiation. More than 20% stromal cells showed immunostaining for Ki67. However no lymphnode metastasis was seen. The patient was discharged and was advised a 6 monthly follow-up. Two years since then, she remains asymptomatic with no clinical evidence of recurrence and a well-healed scar.

Discussion:

Phyllodes tumor are rare biphasic fibro epithelial tumors of the breast accounting for approximately 0.5% of breast neoplasms and affect women in fourth or fifth decade of life.

These tumors are further classified as benign, borderline and malignant based on stromal cellularity, atypia, mitosis and margins.

Malignant phyllodes have a high propensity to metastasize and recur following excision. The common sites of metastasis are lung, liver and bone. [6]

Heterologous differentiation has been reported in malignant phyllodes. Malignant phyllodes with osteosarcomatous differentiation are highly aggressive and extremely rare constituting 1.3% of such cases [6]. It has been hypothesized that malignant change of metaplastic stromal cells in the tumor, result in these neoplasms [7]. Further, the proliferation rate of osteoblastic cells is higher than that of the stromal cells of PT which is compatible which the aggressive behavior of osteosarcoma, a clearly malignant neoplasm. The closest differential of osteosarcomatous variant of PT is metaplastic carcinoma of the breast with bone formation, which has a different behavior and prognosis; hence a meticulous histological survey of the specimen is mandated [8].

Various conflicting theories have been postulated for the pathogenesis of this neoplasm. The first one suggests differentiation from embryonic pluripotent cells. This theory holds good and explains the development of foci of osteosarcomatous differentiation, however, the negativity for epithelial markers in the sarcomatous cells argues against this theory [8].

More than 40 percent of osteogenic sarcomas have a preexisting fibroadenoma or phyllodes tumor, as described in the present case scenario, and we hypothesize that these neoplasms might originate from metaplasia of stromal cells that then undergo malignant transformation. Clinically, PT presents as rapidly enlarging non-tender breast lumps. Mammography reveals dense well- circumscribed or lobulated lesion with areas of calcification, which is confirmed by other imaging techniques such as sonography or MRI scan [9]. The pre operative diagnosis of PT can be made on fine needle aspiration cytology or core biopsy.

The treatment modality for these tumors is mastectomy with complete surgical excision of the tumor and breast tissue. Axillary dissection is warranted only in patients with palpable lymph nodes. The role of chemotherapy and radiotherapy remains controversial [9]. Axillary dissection was done in the present case due to microscopic evidence of infiltrating ductal carcinoma.

To conclude, osteosarcomatous differentiation in malignant phyllodes tumor is a rare occurrence. Early diagnosis and complete surgical resection prevents the recurrence of such tumors and improves patient outcome.

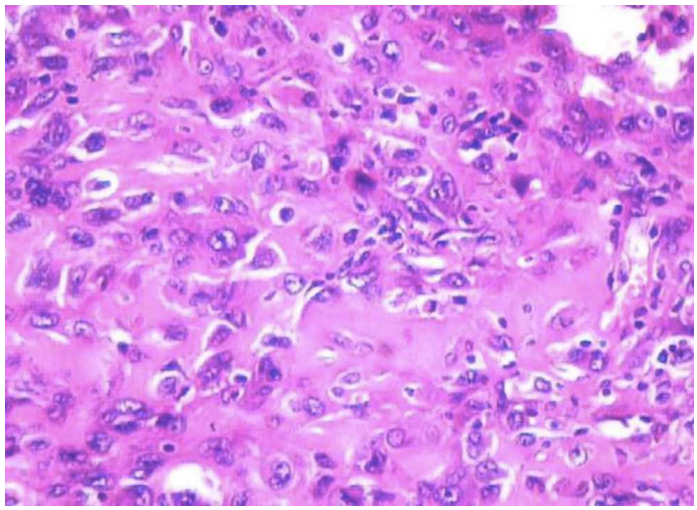

Figure 1: Photomicrograph of phyllodes tumor showing stromal hypercellularity (H&E, X20).

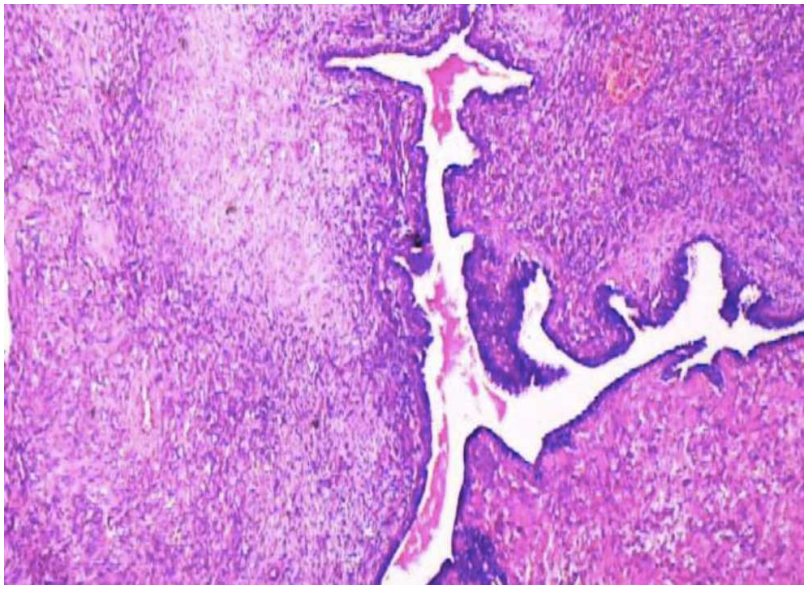

Figure 2: Photomicrograph showing focus of osteosarcomatous differentiation (H&E, X40).

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