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Case Report

Malignant phyllodes tumor with heterologous sarcomatous element: A rare case report

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ABSTRACT

Background: Phyllodes tumors being a rare neoplasm of the breast attributes only 0.3% to 1% of all breast tumors, with an annual incidence of about 2 per million women. Malignant differentiation of phyllodes tumors can occur, resulting in cases of extremely rare heterologous sarcomatous differentiation.
Case Report: We present a rare case of a 63-year-old women with features of malignant phyllodes with sarcomatous (chondrosarcomatous) differentiation.
Conclusion: Morphologically, phyllodes tumors are categorized as benign, borderline and malignant based on the scope of stromal atypia, mitotic activity, stromal overgrowth, and the tumor margins. The presence of heterologous elements in a phyllodes directly upgrades it to a malignant category. A close follow-up with a CT scan is of utmost need to detect the recurrence and metastasis to the earliest.

1. Introduction

Malignant phyllodes tumors (PTs) are rare fibroepithelial malignancies of the breast, comprising of both epithelial and stromal components and account for 0.3% to 0.5% of all breast neoplasms.[1] These tumors commonly occur in females during the 4th or 5th decade of life. The World Health Organization classified phyllodes tumors into benign, borderline, and malignant categories based on the degree of stromal cellular atypia, mitotic activity per 10 high-power fields, degree of stromal overgrowth, tumor necrosis, and margin appearance. Borderline tumors have the greatest tendency for local recurrence. Majority of the phyllodes tumors are benign (35% to 64%), however all forms of phyllodes tumors have malignant potential and can behave like sarcomas with blood-borne metastasis to various organs, commonly the lungs, bone, and abdominal viscera.[2] The remainder of the phyllodes tumors are categorized as borderline and malignant. The risk of local recurrence ranges from 17% in benign PTs to 27% in malignant PTs. Distant metastasis occurs in up to 9.0 – 27.0% of malignant PTs.[3] Heterologous sarcomatous stromal elements, such as, chondrosarcoma, liposarcoma, osteosarcoma, rhabdomyosarcoma, angiosarcoma, and leiomyosarcoma are rarely chanced upon in a malignant Phyllodes tumor.[4] We present a rare case of malignant phyllodes with sarcomatous (chondrosarcomatous) differentiation.

2. Case Report

A 63-year-old female patient presented with a hard lump in the inferior quadrant of the breast for 4 months. Mammography of the left breast revealed mixed glandular and fatty architecture and a well-defined soft tissue mass lesion measuring 6.3X4.1cm, showing smooth margin in the lower central quadrant (BIRADS 3). FNAC was inconclusive and yielded scant cellular material with occasional clusters of benign ductal cells, occasional

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atypical spindle cells and scant stromal fragments. After the necessary investigations and patient's will not to repeat FNAC, she underwent left simple mastectomy under general anesthesia and erector spinal block. The mastectomy specimen was received, and it measured 13.5X9.5X3.5cm. The overlying skin flap measured 11.5X 8.5cm. A horizontal linear scar was noted in the inferior quadrant measuring 8.5 cm. The nipple and areola were within normal limits.

On serial sectioning, a grey-white infiltrating growth with central cystic areas with mucoid secretions measuring 8X8.2X4.7cm was seen involving central quadrant extending to the upper and lower quadrant and to the skin (Figure 1). The deep resection margin was free from tumor grossly.

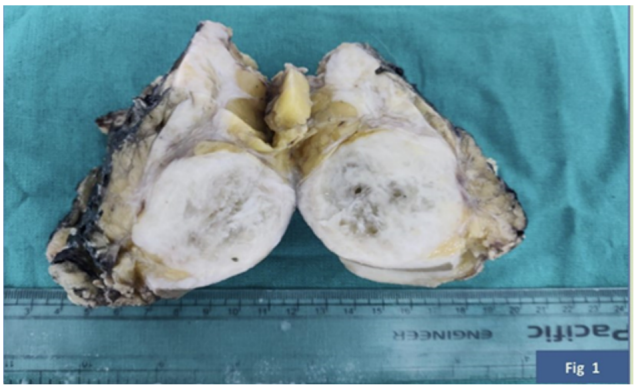

Fig. 1: Showing grey white solid growth with focal cystic and mucoid areas

Microscopic examination revealed a biphasic lesion showing sheets of malignant spindle cells with moderate to high nuclear atypia, increased mitotic activity (MI> 10/10 HPF) and moderate cytoplasm. Stromal overgrowth with infiltrative borders admixed with occasional benign ducts were noted (Figures 2 and 3). In addition, foci of chondrosarcomatous differentiation as evidenced by sheets of atypical and binucleate chondrocytes showing crowding, pleomorphism and hyperchromasia were also seen (Figures 4 and 5). Other features observed were areas of myxoid degeneration and hyalinization. The overlying skin revealed an atrophic epidermis with malignant tumor cells infiltrating in the dermis. The deep resection margin was free of tumor. S-100 showed intense positivity, with diffuse disposal on tumor cells (Figure 6). So based on the clinical features, gross examination, histopathological examination and IHC, a diagnosis of Malignant Phyllodes Tumor with heterologous sarcomatous element (chondrsarcomatous component) was made. The follow-up period was uneventful, and she is living free from disease for the past one year with regular follow –up.

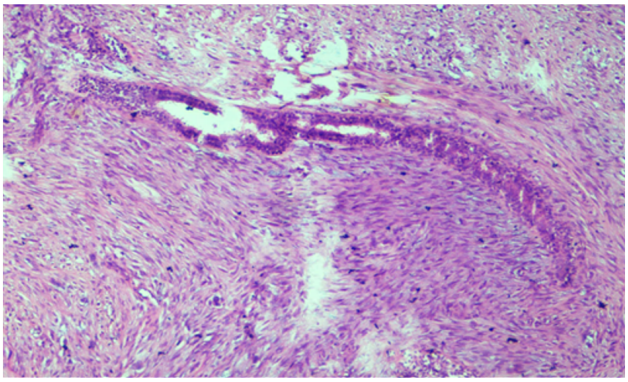

Fig. 2: Showing a biphasic tumor (glandular and stromal components)

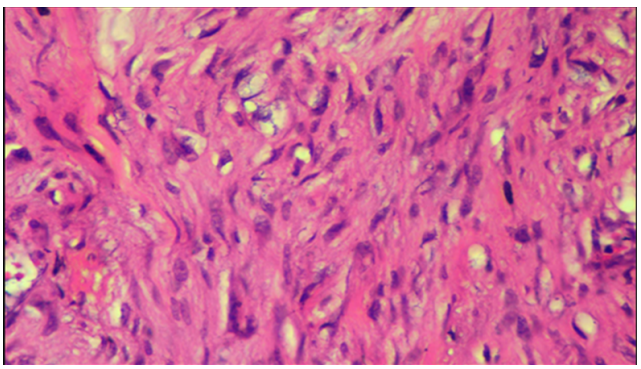

Fig. 3: Showing malignant stromal component

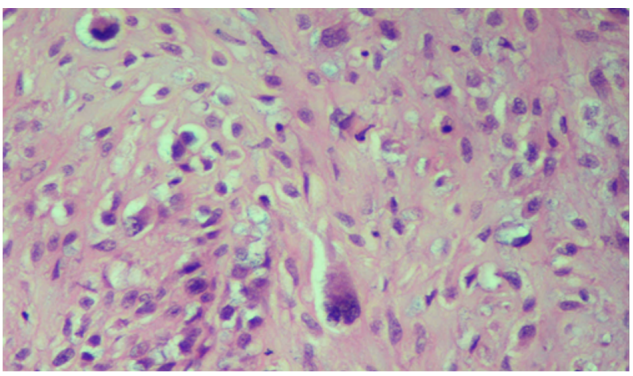

Fig. 4: Showing chondrosarcomatous differentiation with binucleate chondrocytes

3. Discussion

Phyllodes tumors are fairly rare neoplasms of the breast attributing only 0.3% to 1% of all breast tumors. An annual incidence of about 2 per million women has been reported in literature.[5] Morphologically, they are categorized as benign, borderline and malignant based on the scope of stromal atypia, mitotic activity, stromal overgrowth, and the tumor margins. Microscopically, PTs are distinguished by a leaf-

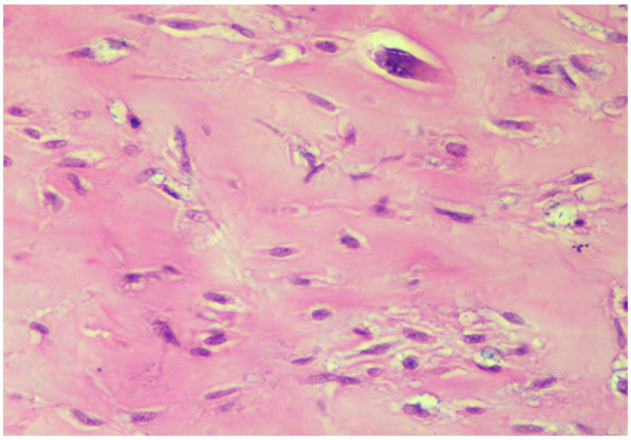

Fig. 5: Showing chondrosarcomatous differentiation with binucleate chondrocytes

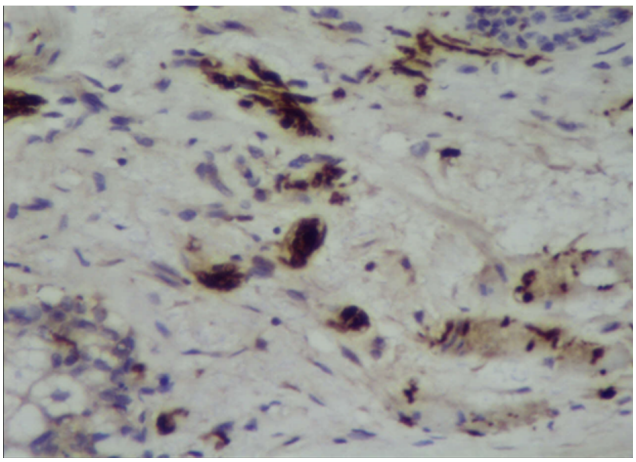

Fig. 6: Showing tumor cells with S-100 positivity

like appearance, generated by projections of hypercellular stroma into epithelium lined cystic spaces. There is presence of dual population of both epithelial and stromal elements, which is necessary for the diagnosis of PT.

Benign PTs can be difficult to distinguish from fibroadenomas while malignant PTs can grow quickly and metastasize early. However, the characteristic histological features are related to the stroma and PTs are differentiated from fibroadenomas by the presence of marked stromal overgrowth and hypercellularity.

A variety of associated malignancies are seen to arise from PTs, with their dual population of cells. The stromal cells can illustrate sarcomatous differentiation while the epithelial component can transmute into malignant with DCIS/LCIS or invasive carcinoma. There have also been case reports of two distinctly separate lesions of PT and primary sarcoma or PT and carcinoma.[1] The presence of heterologous elements in a phyllodes directly upgrades it to a malignant category. These tumors with heterologous sarcomatous differentiation which comprise of liposarcoma, angiosarcoma, chondrosarcoma, rhabdomyosarcoma or osteosarcoma, are rare, accounting for 1.3% of all phyllodes tumors.[4] Recent data suggest that malignant PT cells exhibit mesenchymal stem cell characteristics and can be induced or spontaneously differentiated into other lineages, consistent with previous reports on the heterogenic differentiation of PT.[6] The differential diagnosis includes tumors of the breast showing bone and cartilaginous differentiation which include Intraductal papilloma with stromal metaplasia, cystosarcoma phyllodes, stromal sarcoma, and adenocarcinoma with metaplasia.[7]

Surgical treatment is generally the treatment of choice for phyllodes tumors, regardless of its histological subtype. In isolated PTs, wide local excision with margins of 1 cm is recommended with minimal evidence for chemotherapy or hormone therapy.[1] However, it is suggested by some authors to use adjuvant therapy in PTs greater than 5 cm with more than 20 mitoses per 10 high power fields. This is because these tumors may have a higher local recurrence rate, and hence adjuvant radiotherapy was offered in the present case. Lymph node involvement is rarely seen in phyllodes tumors and routine axillary lymph node dissection is thus often avoided. The most common path is the hematogenous spread, which occurs mostly in the lungs, pleura, and bones.[8]

The prognosis for malignant phyllodes tumors is poor and the role of various treatment modalities is not clearly defined with local recurrence rates ranging from 10 to 40% (average 15%) and distant metastases occurring in 10% of all phyllodes tumors, and up to 20% of malignant phyllodes tumors.[9]

4. Conclusion

We conclude by saying that Phyllodes tumors with heterologous sarcomatous differentiation are extremely rare with early recurrence and metastatic potential. A close follow-up with a CT scan is of utmost need to detect the recurrence and metastasis to the earliest. Role of adjuvant radiotherapy and chemotherapy although not clearly defined, but they may be administered in patients with high-grade tumors (as in our case), positive surgical margins, or postoperative recurrence.

5. Source of Funding

None.

6. Conflict of Interest

None.

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