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Case Report

Malignant phylloides with heterologous elements: A case report

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ABSTRACT

Phyllodes tumor of the breast is an uncommon biphasic tumor, occurring mainly in females belonging to the age group of 42 to 45 years. Malignant phyllodes tumor is a rare neoplasm that accounts for 10 to 20% of total phyllodes tumors. Histologically, phyllodes tumors are featured by leaf like projections of hypercellular stroma into the cystic or cleft-like spaces being lined by epithelium. Presence of stromal elements like fibrosarcomatous and heterologous sarcomatous characters are associated with an increased risk of malignant change. Here, we report a rare case of a 50yrs old female patient, who presented with swiftly growing left breast mass that was diagnosed as a case of malignant phyllodes tumor showing heterologous elements in form of chondroid, liposarcomatous, rhabdoid and fibrosarcoma, with marked proliferation of stromal component. The patient was subjected to modified radical mastectomy.

1. Introduction

Phyllodes tumor of the breast is known as an uncommon biphasic tumor with stromal and epithelial components.[1] They mainly occur in females belonging to the age group of 42 to 45 years. They are usually featured as a fast growing big palpable mass.[2] They had their origin from peri-ductal stroma and are categorised as malignant, borderline or benign, based on their histological characteristics like cellularity, overgrowth and atypia of stromal tissues, mitotic rate, and borders of tumor.[3] Most of the phyllodes tumors of the breast accounts for 2.5% of total fibroepithelial tumors. They are mainly benign, estimating for 60 to 75% of total phyllodes tumors. Malignant phyllodes tumor is a rare neoplasm that accounts for 10 to 20% of total phyllodes tumors and 0.18% of total malignant breast cancers. But it has been observed that all phyllodes tumors have the capability to turn into malignant tumors.[4] These tumors are generally solitary in nature but sometimes they can be found linked with other breast cancers. They can also be seen co-existing with a clearly separated malignant cancer in both ipsilateral or contralateral breasts.

Histologically, phyllodes tumors are featured by leaf like projections of hypercellular stroma into the cystic or cleft-like spaces being lined by epithelium. Many variations have been observed in the histological patterns of different tumors. These can vary from being resembling fibroadenomas, (having raised mitotic activity and stromal cellularity) to soft tissue sarcoma (with diffuse overgrowth of stromal cells which are highly pleomorphic) and even showing a range of appearances midway between these two extreme conditions. Stromal elements like fibrosarcomatous and heterologous sarcomatous characters are associated with an increased risk of malignant change. Heterologous variations like osteosarcoma, chondrosarcoma, liposarcoma, angiosarcoma, and rhabdomyosarcoma, are rarely seen.[5–7] Differential

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diagnosis of fibromatosis, breast cancer, myxoid sarcoma, angiosarcoma, myofibroma, juvenile fibroadenoma was considered.[8]Little information iavailable about imaging findings of cases cited in the literature.

In this present case report, we present a case of malignant phyllodes tumor with its pathological findings. This rare case was characterized by stromal overgrowth, chondrosarcomatous differentiation, having fibrosarcoma, liposarcoma and rhabdoid components.

2. Case Report

A 50 years old female presented with a left breast mass that was initially small and had rapidly increased in its size over a period of 4 years. The swelling was associated with pain, with no history of fever, weight loss, nipple discharge, anorexia, bone pain and loss of consciousness. Past medical history was not suggestive of tuberculosis, diabetes mellitus, asthma, hypertension or any chronic disease. She had no family history of breast cancer, and did not take hormone replacement drugs or oral contraceptives. She didn't reported with any history of drug allergy. On general examination, no pallor, clubbing, icterus, cyanosis, lymphadenopathy, and pedal edema was detected. Vitals were recorded to be normal. No abnormality was detected on systemic examination in relation to CNS, CVS, and respiratory system.

Physical examination of breast revealed that left breast was bulky with nipple at lower level than right breast. Swelling in left breast was involving the whole breast. Skin over the swelling was shiny, tense with tortuous veins. No signs and symptoms of nipple discharge, venous engorgement, nipple retraction, and puckering were observed. On palpation, a non tender lump involving whole of the left breast was found with no local rise in temperature. Lump was movable with respect to the skin and chest wall. Axillary and supraclavicular lymphnodes were not palpable.

Breast ultrasonography (US) detected an ill defined heterogeneous solid and cystic lesion in left breast. The lesion was characterised by internal calcification, and spiculous margins, measuring $20 \times 15 \times 10$ cm. Visualised portion of left chest wall shows no invasion. The mass was categorized as Breast Imaging Reporting and Data System (BI-RADS) 5. Tru Cut biopsy showed cellular stromal component with extensive hyalanization and moderate stromal atypia, suggestive of boderline phyllodes tumor. The patient underwent left modified radical mastectomy without dissection of axillary pad of fat measuring 19 x 13 x 9 cm. Drain was removed on 2^{nd} post operative day and patient was allowed for oral intake, that she tolerated well. Patient was then discharged under satisfactory conditions. Patient was recalled for follow up visit after 10days.

Macroscopic evaluation of mastectomy specimens revealed the skin surface with nodule measuring 3 x 3 cm, with no scar. The serial section of the tumor revealed a well-circumscribed, fleshy, nodular, solid white growth with hemorrhage, cystic area and no necrosis. The growth measures 0.8cm from superior margin, 0.2cm approaching medial margin, 3cm from inferior margin, almost approaching 0.3cm from lateral margin and <0.1cm from base. No lymph nodes were identified. Tumor size measured around 19 x 10 x 9cm.

Immunohistochemical stains were used to identify various elements; fibrosarcoma was Vimentin positive, rhabdoid cells were Desmin positive, Chondrosarcoma was s100 positive and special stai, Oil red O was used to identify adipocytes.elements in form of chondroid (Figure 1), liposarcomatous (Figures 2, 3 and 4) like elements. Section showed a marked component along with interspersed benign duct Figure 1. Stroma is higAreas of necrosis were observed with margins being free of tumor, and no perineural and lymphovas

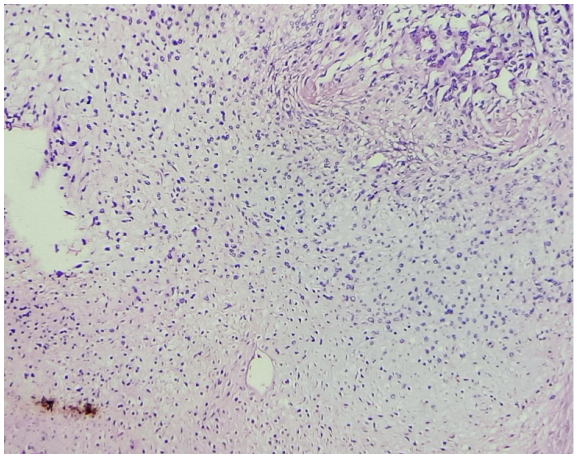

Fig. 1: Malignant phyllodes tumor displaying chondrosarcoma component (20x magnification)

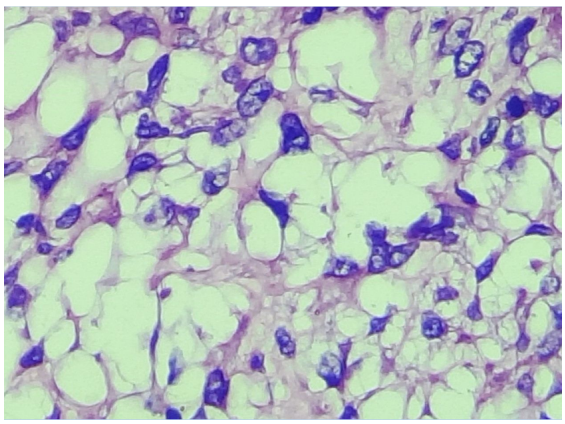

Fig. 2: Malignant phyllodes tumor displaying liposarcomatous component (20x magnification).

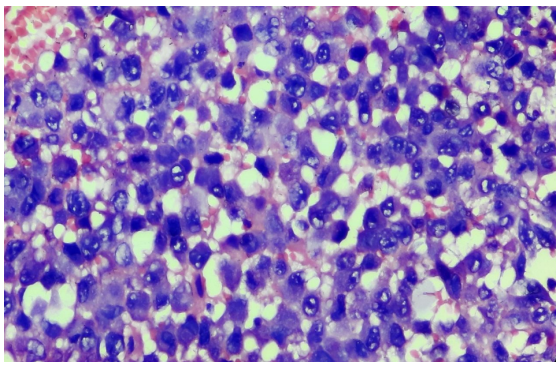

Fig. 3: Malignant phyllodes tumor displaying rhabdoid component (20x magnification).

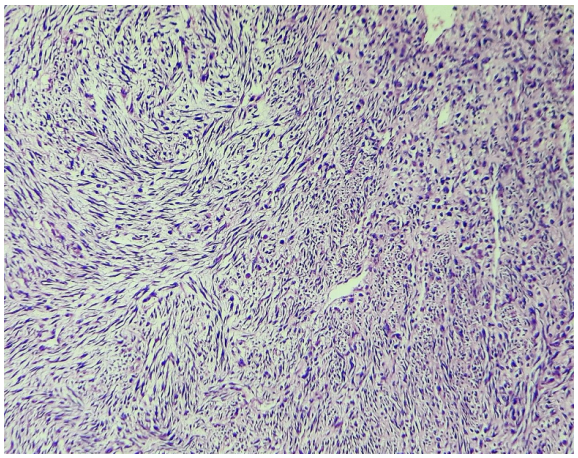

Fig. 4: Malignant phyllodes tumor displaying fibrosarcoma component (20x magnification).

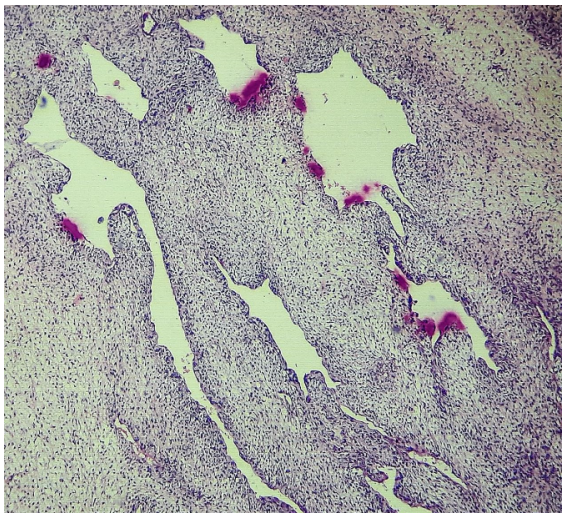

Fig. 5: Malignant phyllodes tumor displaying proliferation of stromal component (10x magnification)

3. Discussion

Phyllodes tumors are type of fibroepithelial neoplasms which were first reported in 1838 by Muller as Cytosarcoma phyllodes.[9] These tumors are characterised by a variety range of clinical, cytological and histological features, showing cellular stromal and epithelial components.[10]Based on five different histological characters: mitotic activity, stromal cellularity, tumor margins, stromal atypia, and stromal overgrowth, World Health Organization has classified phyllodes tumors into malignant, borderline, and benign types.[1] Benign phyllodes tumor was found to occur in younger age group as compared to borderline and malignant phyllodes tumors.

When the tumor shows increased mitotic activity (≥10 per 10 high power fields), marked stromal hypercellularity and atypia, permeative borders, and stromal overgrowth, it is diagnosed as malignant phyllodes tumor. It also reflects the occurrence of malignant heterologous sarcomatous elements like chondrosarcoma, liposarcoma, or osteosarcoma.[7] In present case report we also diagnosed the case to be malignant phyllodes tumor, as we observed the presence of leaf like patterns with areas of heterogenous elements in form of chondroid, liposarcomatous, rhabdoid and fibrosarcoma, with marked proliferation of stromal component. US characteristics like presence of cystic mass, increased vascularity, and cleft-like rounded cystic space reflect the presence of phyllodes tumor.[11] It has been observed that majority of recurrence is similar to the initial tumor histologically, however malignant transformation with heterologous differentiation is rare but it has been described.Wiratkapun C et al.,[12] we also observed ill defined heterogeneous solid and cystic lesion in left breast. The lesion was characterised by internal calcification, and spiculous margins.

The definitive management for phyllodes tumor is mastectomy or wide surgical excision of affected area with about 1-2 cm of negative margins based on the tumor and breast size. It has been advocated that benign phyllodes tumors hold a good prognosis after surgical intervention, but malignant phyllodes tumors are clinically aggressive, with high rate of local recurrence (23-30%) and distant metastasis rate being 20%.[2] Characteristics like severe stromal overgrowth, nuclear pleomorphism, infiltrating tumor margins, and high rate of mitosis are known the predictors of metastasis.[10] Various studies revealed that the occurrence of heterologous elements were mainly linked with causation of distant metastasis having a poor outcome.[1–5] Lymphatic spread is uncommon in such tumors, so routine dissection of axillary lymph node is not generally recommended. In our case, biopsy was done sparing lymph nodes, as axillary and supraclavicular lymphnodes were not palpable.

Limited studies are available till now in the present literature to reveal the prognosis of phyllodes tumors

having heterologous sarcomatous growth and to guide about the role of adjuvant therapy in their treatment. Thus in future various prospective studies and clinical trials should be conducted to generate high-level data regarding management of malignant phyllodes tumors.

4. Conclusion

The present article reports a rare case of malignant phyllodes tumor diagnosed in a 50yrs old female patient with heterologous elements in form of chondroid, liposarcomatous, rhabdoid and fibrosarcoma, with marked proliferation of stromal component. As limited scientific data is available regarding the treatment of such tumors, various clinical trials and prospective multicenter studies with long-term follow up are required to develop treatment guidelines for phyllodes tumor having heterologous sarcomatous growth.

5. Source of Funding

None.

6. Conflict of Interest

None.

References

1. Ko SY. Malignant phyllodes tumor of the breast with heterologous osteosarcoma and chondrosarcomatous differentiation: A rare case report with imaging findings. *Radiol Case Rep*. 2023;18(5):1982–8.
2. Warrier S, Hwang SY, Gibbings K, Carmalt H, O'Toole S. Phyllodes tumour with heterologous sarcomatous differentiation: Case series with literature review. *Int J Surg Case Rep*. 2015;11:91–4.
3. Goto W, Kashiwagi S, Takada K, Asano Y, Morisaki T, Noda S, et al. A case of a malignant phyllodes tumor that was difficult to distinguish from stromal sarcoma. *Gan To Kagaku Ryoho*. 2018;45(13):2429–31.
4. Vorherr H, Vorherr UF, Kutvirt DM, Key CR. Cystosarcoma phyllodes: epidemiology, pathohistology, pathobiology, diagnosis, therapy, and survival. *Arch Gynecol*. 1985;236(3):173–81.
5. Tan PH, Ellis I, Allison K, Brogi E, Fox SB, Lakhani S, et al. The 2019 World Health Organization classification of tumours of the breast. *Histopathology*. 2020;77(2):181–5.
6. Nayak M, Patra S, Mishra P, Sahoo N, Sasmal PK, Mishra TS, et al. Malignant phyllodes tumor with heterologous differentiation: Clinicopathological spectrum of nine cases in a tertiary care institute in Eastern India. *Indian J Pathol Microbiol*. 2017;60(3):371–6. doi:10.4103/IJPM.IJPM_426_16.
7. Koh VCY, Thike AA, Nasir NDM, Yip GWC, Bay BH, Tan PH, et al. Size and heterologous elements predict metastases in malignant phyllodes tumours of the breast. *Virchows Arch*. 2018;472(4):615–21.
8. Jagtap SV, Nagar V, Bhosale SJ, Nagar D, Jagtap SS. Nodular Fasciitis Of The Breast - An Uncommon Benign Reactive Proliferative Lesion. *IP Arch Cytol Histopathology Res*. 2021;6(4):291–4.
9. Pornchai S, Chirappapha P, Pipatsakulroj W, Lertsithichai P, Vassanasiri W, Sitathanee C, et al. Malignant transformation of phyllodes tumor: a case report and review of literature. *Clin Case Rep*. 2018;6(4):678–85.
10. Soni N, Aly NT, Vidholia A, Policeni F. Not every fat-containing lesion on mammogram is a benign finding: case report and review of breast malignant phyllodes tumor with heterologous liposarcomatous differentiation. *J Clin Imaging Sci*. 2019;9:10. doi:10.25259/JCIS_83_18.
11. Kalambo M, Adrada BE, Adeyefa MM, Krishnamurthy S, Hess K, Carkaci S, et al. Phyllodes tumor of the breast: ultrasound-pathology correlation. *AJR Am J Roentgenol*. 2018;210(4):173–9.
12. Wiratkapun C, Piyapan P, Lertsithichai P, Larbcharoensub N. Fibroadenoma versus phyllodes tumor: distinguishing factors in patients diagnosed with fibroepithelial lesions after a core needle biopsy. *Diagn Interv Radiol*. 2014;20(1):27–33.

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