

Primary Chondrosarcoma of The Breast: A Rare Case Presentation

Anshu Jain, Veena Maheshwari, Shagufta Qadri, Divya Rabindranath*, Nishat Afroz

Department of Pathology, J.N. Medical College, A.M.U. Aligarh, India

Keywords: *Breast, Chondrosarcoma, Cytology, Primary*

ABSTRACT

Pure sarcomas are very uncommon tumors of the breast. Only ten case reports of primary chondrosarcoma of breast have been published in literature till date. We report this rare case highlighting the cytological features which helped in a preoperative diagnosis of this tumor. A 40 year old lady presented with lump and pain in right breast. She did not have any other significant findings. Fine needle aspiration cytology yielded abundant material, comprising of benign ductal epithelium along with malignant cartilaginous component. Subsequent histopathology and immunohistochemistry confirmed the cytological diagnosis of primary chondrosarcoma of breast. Primary chondrosarcoma of breast, even though a rare entity, should be kept in the differential diagnosis of breast tumours exhibiting chondrosarcomatous areas.

***Corresponding author:**

Dr. Divya Rabindranath, Department of Pathology, J.N. Medical College, A.M.U., Aligarh-202002. Uttar Pradesh, India.

Phone: +91- 7895683197

E-mail: divy30@hotmail.com



Introduction

Primary pure sarcomas of breast represent only 0.5% of breast malignancies.^[1] The differential diagnosis includes metaplastic breast carcinoma with chondrosarcomatous differentiation and cystosarcoma phyllodes.^[2] It is important that they are recognized as a separate entity from the more common breast carcinomas and the difference in behavior of these two tumors is taken into account when planning therapy.^[1]

Case Report(S)

A 40 year old Asian female presented with right breast lump since one year. She had developed pain in the lump since seven days. There was no history of lump or pain anywhere else in the body. There was no family history of breast cancer and patient was multiparous. On examination, there was an 8 x 8 cms hard lump in the right breast. Overlying skin was unremarkable, apart from few prominent veins. Lump was not fixed to underlying chest wall. No axillary or supraclavicular lymphadenopathy was noted. Left breast was normal. Rest of the systemic and general examination was unremarkable. Her routine laboratory investigations were normal. A chest radiograph was also normal with normal ribs. A provisional clinical diagnosis of carcinoma breast was made and patient was advised fine needle aspiration cytology (FNAC). Multiple site sampling from the mass was done. The FNAC smears were highly cellular and showed sheets of benign ductal cells along with loose aggregates and scattered population of moderate to markedly atypical cells against a chondroid background (Figures 1a, 1b). At places typical chondroblastic morphology was also appreciable (Figure 1c). Only occasional stromal fragments and few mitosis were noted. This led to a differential diagnosis of chondrosarcoma of breast. A diligent search for any chondrosarcomatous lesion in bones was made,

and none found. Patient underwent right modified radical mastectomy with axillary lymph node dissection. On grossing, the mastectomy specimen measured 15x9x4 cm with overlying ellipse of skin measuring 11x8 cm. Cut sections showed a tumor 10x7x6 cm in size. It was whitish, friable with foci of cystic change along with chondroid areas (Figure 2a). Tumor distance from all margins was ≥ 1.0 cm. 10 lymph nodes were dissected out, none of which seemed involved grossly. Extensive sampling for histopathological examination was done. Microscopically the tumor was composed entirely of lobules of chondroid material with moderate to markedly atypical chondrocytes (Figures 2b, 2c). These lobules were separated by fibrotic stroma. The tumor showed involvement of adipose tissue of breast. Benign ductal component could be seen at the periphery of tumor. Immunohistochemically, the tumor cells were S 100 positive (Figure 3a); Pan Cytokeratin, Estrogen Receptor, Progesterone Receptor, & Her2 neu negative (Figures 3b, 3c, 3d, 3e). Skin, all margins & the lymphnodes were free from tumor. A final diagnosis of primary chondrosarcoma of breast was made and as it was fully excised, patient was kept under observation with regular followup. She is doing well six months after surgery.

Discussion

Most pathologic lesions in the breast arise from the epithelium. The stroma, however, may also give rise to neoplasms; in general, these are similar in appearance to mesenchymal lesions seen at other sites. Pure sarcomas, without an associated epithelial component, occasionally occur in the breast and should be distinguished from metaplastic carcinoma and malignant phyllodes tumor that has outgrown its epithelial component.^[3]

Fewer than 150 cases of primary osteosarcoma of breast have been reported in the medical literature. Primary

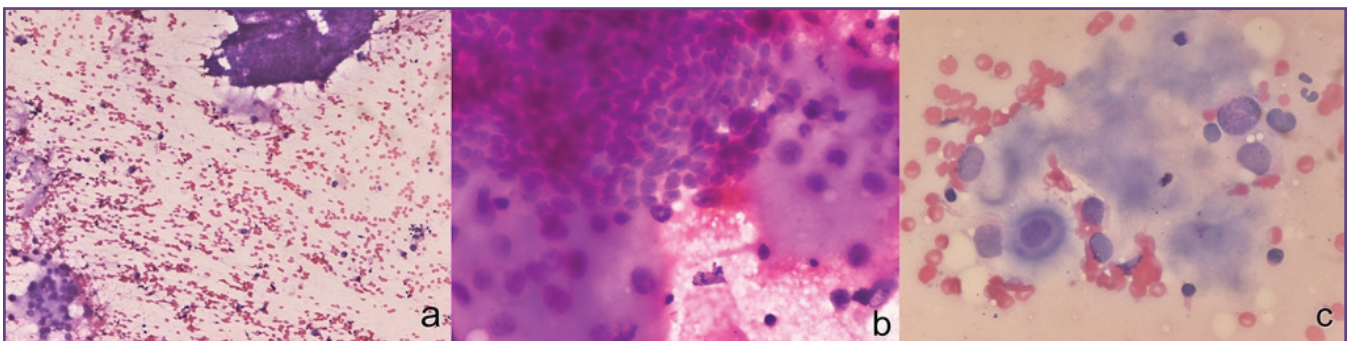


Fig. 1 [a]: Photomicrograph showing branching sheet of benign ductal cells (upper half) and malignant chondroid component (lower half). (H&E, x100); **[b]** Benign ductal cells in close proximity with atypical malignant cells with a chondroid background. (H&E, x400); **[c]** Typical chondroblast with surrounding lacunar space and background chondroid material. Adjacent atypical cells can be confused with malignant ductal component, and erroneously diagnosed as ductal carcinoma if attention to chondroblasts is not given. (H&E, x400).

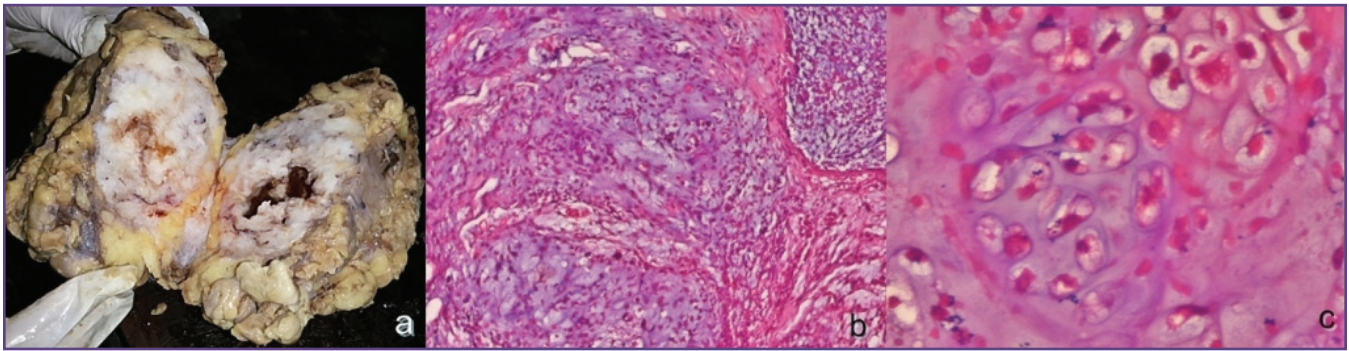


Fig. 2 [a]: Gross photograph of cut section of tumor showing a whitish and friable appearance; **[b]** Lobules of chondroid material with atypical chondroblasts. (H&E, x100); **[c]** Higher power showing atypical nuclear morphology as well as hypercellularity. (H&E, x400).

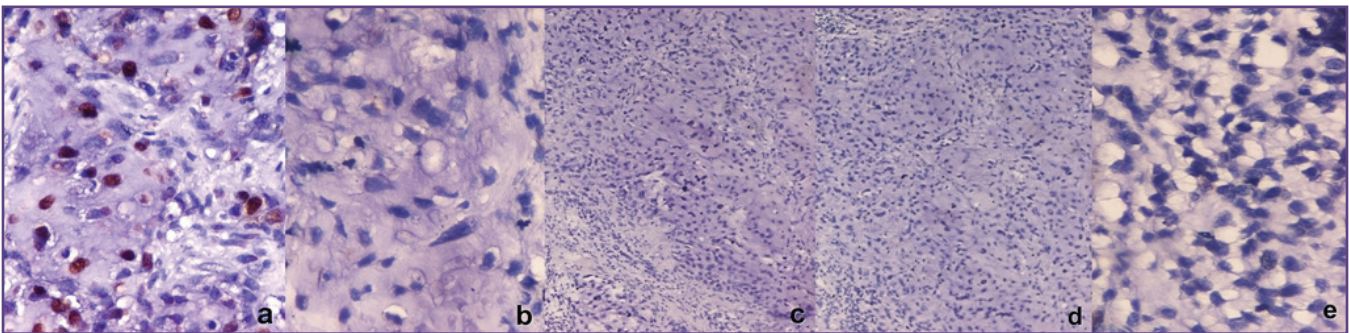


Fig. 3: Immunohistochemistry of tumor cells showing [a] S100 positivity (x400); while negativity for [b] Pan Cytokeratin (x400), [c] ER (x100), [d] PR (x100), & [e] Her2 neu (x400).

chondrosarcoma is even rarer and publications are limited to isolated case reports or few cases in series encompassing a heterogeneous group of mammary sarcomas.^[4] The existence and histogenesis of primary mammary matrix producing (MP) sarcomas, namely osteosarcoma and chondrosarcoma, remains controversial. It is still unidentified whether these represent the end stage of trans-differentiation towards osteochondroid lineage with loss of residual epithelial components (carcinoma); or these are tumors arising from totipotent mesenchymal cells of the breast stroma (sarcoma).^[4]

A preoperative clinical and cytological diagnosis in such cases is usually not reached, both due to marked similarity in clinical behaviour to infiltrating ductal carcinoma and low index of suspicion.^[2] The cytological findings of primary breast chondrosarcoma have never been elucidated before. In our case, the presence of large amount of benign ductal component indicated a non-epithelial nature of the malignancy and raised the strong suspicion thereby confirming the retrospective diagnosis of primary chondrosarcoma on cytological basis. The malignant chondroid component can be mistaken for ductal carcinoma, however one should pay attention to the

background matrix material and typical chondroblastic morphology present focally. Only occasional stromal fragments were seen which is non-favourable for phyllodes tumor, although it is very difficult to distinguish only by cytology.

Even on histopathology, it is sometimes very difficult to distinguish a primary chondrosarcoma from metaplastic breast carcinoma and malignant phyllodes tumor, if the chondrosarcomatous component is predominant. In such cases wide sampling is important in order to rule out any foci of in situ or invasive epithelial malignancy. Any such foci would lead to a diagnosis of metaplastic breast carcinoma.^[3]

In the occasional case reports, these tumors have usually been found to be large in size, occur in women more than 40 years old, and usually do not invade the overlying skin.^[5] Regional lymphadenopathy is expected in 14-29% of these cases, most of which are reactive hyperplasias.^[6] Microscopically the tumor shows chondroid areas with cellular atypia and pleomorphism.^[2] Differentiation from metaplastic carcinoma is possible by absence of direct transition between carcinomatous and mesenchymal component in the former. Further the sarcoma like

elements in metaplastic carcinoma though acquire vimentin positivity, still retain epithelial markers.^[7]

Differentiation from malignant cystosarcoma phyllodes with predominant chondrosarcomatoid component can be extremely difficult. It has been mentioned that benign ductal elements interspersed- among sarcomatoid areas should be taken as evidence of former. Most mammary tumours with areas of chondroid metaplasia show benign histological appearance.^[2] Another important differential diagnosis that should be considered in such cases is primary sarcoma of the chest wall metastasizing to the breast. The only method to rule out this possibility is thorough search for lesions in chest wall preoperatively through imaging; and during surgery.

Surgery remains the treatment of choice for most sarcomas. The role of chemotherapy and radiotherapy is not yet established because of the limited number of cases reported so far.^[1]

Conclusion

In conclusion, a preoperative diagnosis in a case of primary chondrosarcoma of the breast is important as it would surely negate the need for un-necessary preoperative chemotherapy to the patient. This is important because chondrosarcomas are primarily managed surgically, and generally have good prognosis once completely excised as compared to the ductal carcinomas.

Acknowledgements

Nil

Funding

None

Competing Interests

None declared

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