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Osteogenic Sarcoma of the Breast- A Rare Case of Extraosseous Tumour

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Abstract: *Breast cancer is the commonest cancer that afflicts females worldwide. Of all the cancers of the breast, carcinoma forms the bulk while breast sarcomas are negligible. Extraskeletal osteogenic sarcomas [OS] account for less than 1% of soft tissue sarcomas.When this tumour develops in the breast, it originates either from normal breast tissue de novo, or as metaplastic differentiation of a primary benign or malignant breast lesion. Secondary deposits from a primary bone sarcoma occur only rarely. We present the case of a 27 year old young woman who was diagnosed with an osteogenic sarcoma of the breast. We have performed a thorough literature review and would like to use this case to highlight several details of this unusual tumour.*
Key words: *osteogenic sarcoma, breast, metaplastic differentiation*

I. Introduction

Primary sarcomas of the breast are extremely rare and make up less than 0.1% of all malignant tumours of the breast . Fewer than 150 cases of primary osteosarcoma of breast have been reported in the medical literature in the last 50 years[1]. Extra-skeletal osteosarcoma has been documented in many tissues of the body including the thyroid gland, kidney, bladder, colon, heart, testis, penis, gall bladder and the cerebellum . Osteogenic sarcomas of the breast either arising primarily in the breast or as secondary deposits from primary bone sarcomas occur in very rare cases. Sarcoma of breast is usually of the spindle cell variety. Most reports in the literature are limited to presentation of single cases but Silver and Tavassoli in 1998, have reported a series of 50 cases of primary osteogenic sarcoma of the breast, focusing mainly on the clinicopathological findings of this rare entity[2] . This paper reports the case of a young woman who presented with recurrent left breast lump which was clinically diagnosed as carcinoma but turned out to be osteogenic sarcoma arising from the breast.

II. Case Report

A 27 year old female presented with a history of a lump in left breast for past two months gradually increasing in size.There was no history of nipple discharge, bleeding or any other symptom. On examination a hard lump in upper outer quadrant was palpable. No evidence of peau de' orange, skin, chest, ribs or nodal involvement neither past radiotherapy nor family history was found. The ultrasound of abdomen and pelvis, chest x- ray, bone scan, haematological and biochemical parameters were within normal limits. Mammography disclosed well- demarcated mass which was partially calcified. Other breast was normal on examination. Fine needle aspiration cytology was positive for malignant cells(fig. 1, 2).

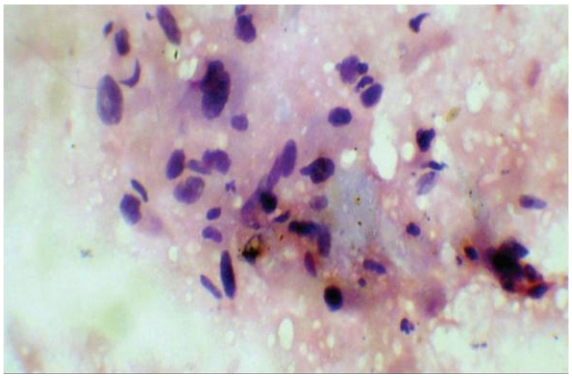

fig. 1

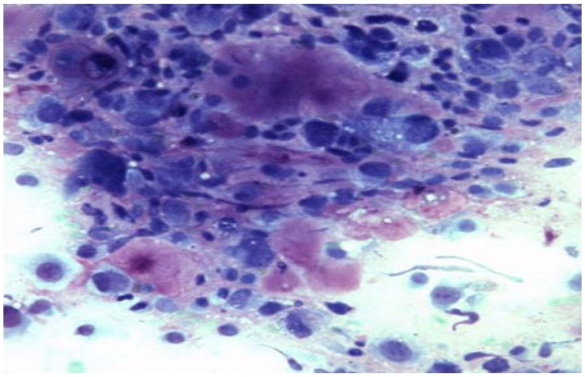

fig. 2

Photomicrographs shows spindle cells with nuclear atypia and pleomorphism. Fragments of malignant mesenchymal elememts seen.(H&E 40X)

She underwent a modified radical mastectomy. The gross specimen was partly well- circumscribed, irregular oval and firm total measuring 10x8x7cm. On cut surface whole of the breast was replaced by solid, fleshy, greyish white mass with hemorrhage and cystic necrotic areas(Fig. 3).

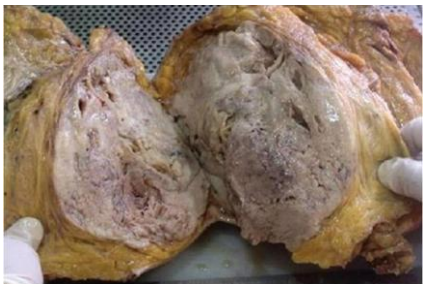

Fig. 3 : Grossly, partly well- circumscribed mass.
Cut surface shows hemorrhagic and necrotic areas.

Twelve axillary lymph nodes were excised. The mass had no involvement of the underlying ribs or the overlying epidermis. Histopathologically, the tumor was essentially well circumscribed. The large pleomorphic epithelioid tumor cells had a syncytial arrangement and were entrapped in the coarsely lacelike osteoid(fig. 4, 5).

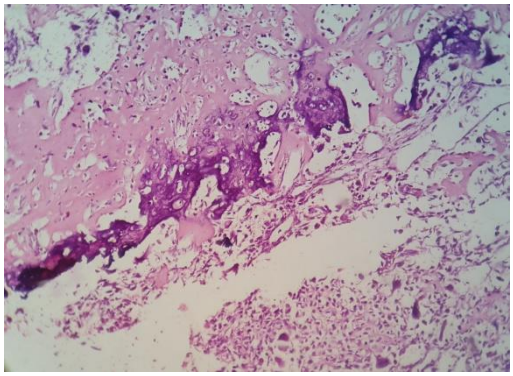

fig. 4

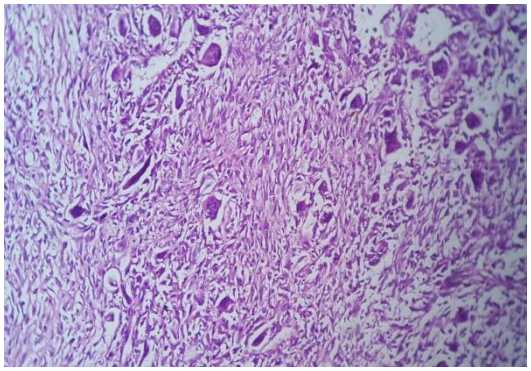

fig. 5

Low power shows discohesive pleomorphic tumour cells with osteoid formation(H&E 10X)

Some areas of the tumor demonstrated a dominant population of spindle-shaped neoplastic cells arranged in a vaguely storiform pattern with finely filigree intercellular osteoid matrix . Bizarre anaplastic tumor giant cells and osteoblast-like tumor cells containing diverse large nuclei, prominent nucleoli and neoplastic osteoid were also present . Few mitotic figures were easily recognized . Multifocal areas of necrosis and hemorrhage within the tumor were seen(fig 6, 7).

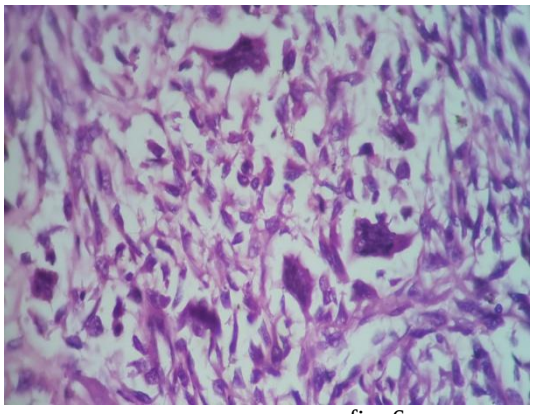

fig. 6

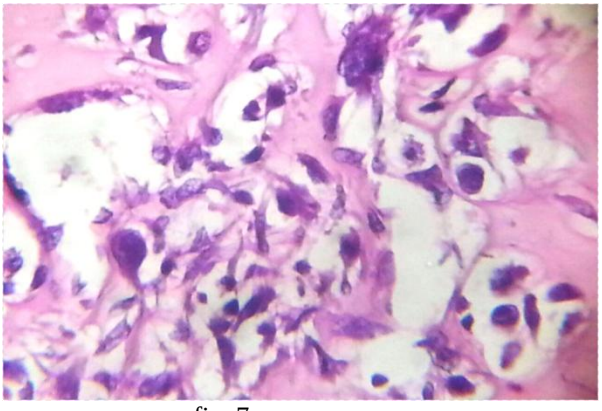

fig. 7

High power shows spindled shaped neoplastic cells and bizarre anaplastic tumour giant cells. Few mitotic figures also seen.(H&E 40X)

There was no histological evidence of an epithelial or a carcinomatous component despite extensive sampling of the tumor. Additionally, a pre-existing phyllodes tumor was not present in any of the examined sections. Immunohistochemical studies displayed diffuse positivity of the neoplastic mesenchymal cells for vimentin , S100 and CD68. The tumor cells had no immunoreactivity for pan-cytokeratin (AE1/AE3), CK7, carcinoembryonic antigen (CEA), epithelial membrane antigen (EMA), high-molecular-weight cytokeratin 34BetaE12, smooth muscle actin (SMA), CD117, CD34, HMB-45, myoglobin, desmin and CD99. The tumor cells are also negative for HER-2/neu oncoprotein as well as estrogen and progesterone receptors. The immunohistochemical studies showed no evidence of epithelial differentiation. According to the above pathological findings, a primary osteoblastic osteosarcoma was diagnosed. The sections from axillary lymph nodes revealed no evidence of tumor metastasis.

III. Discussion

Primary mammary osteosarcoma is an exceptionally rare tumor and represents 12.5% of mammary sarcomas [3]. The criteria required for the diagnosis of a primary osteosarcoma of the breast include (i) demonstration of neoplastic osteoid and neoplastic osteoblasts, (ii) the exclusion of primary bony origin, and (iii) the absence of neoplastic epithelial component that may undergo metaplasia.[4]

Primary mammary osteosarcoma usually occurs in middle-aged or elderly women [5]. The histogenesis of primary osteosarcoma of the breast is not clear, but an origin from totipotent mesenchymal cells of the breast stroma or a transformation from a pre-existing fibroadenoma or phyllodes tumor has been suggested . Mammographically, these tumors often present as a well-circumscribed dense lesion within the breast parenchyma with focal or extensive coarse calcifications[3]. Metastasis is haematogenous to the lungs, bone, skin and brain. Simple mastectomy is the treatment of choice because tumour spread is haematogenous and not via the lymphatic system . Adjuvant chemotherapy may be beneficial but its effect on long-term survival has not been proven . It is an aggressive tumour and carries a worse prognosis than other breast malignancies with reported survival rates of 1–2 years even with treatment.[5]

Osteosarcoma of the breast, similar to other extraskeletal osteosarcomas, may have a broad primary osteosarcomas of the breast are osteoblastic, osteoclastic and fibroblastic subtypes [3].

The diagnosis of metaplastic mammary carcinoma should be excluded before primary breast osteosarcoma is diagnosed, however this was excluded because of negative epithelial markers on immunohistochemical staining. Metaplastic carcinoma is recognised either by the presence of carcinomatous compenent on H&E staining or by cytokeratin immunoreactivity of the neoplastic spindle cells . Secondly, Phyllodes tumour in any sarcoma has, a predominantly fibroblastic component[6]. However, our case had characteristic osteoid formation and osteoclastic giant cells and it is possible to say with conclusively that our case is an osteosarcoma .Treatment for the localized disease should include complete surgical removal of the tumor with an adequate margin. Axillary lymph node dissection is not indicated because axillary node involvement is exceptional[6].

IV. Conclusion

To conclude primary mammary osteosarcoma is an exceptionally rare and highly aggressive neoplasm. This tumor should be distinguished from metaplastic carcinoma or malignant phyllodes tumor with heterologous osseous differentiation because it has different biological behaviors and requires different treatment approaches. Targeted specific therapies, perhaps biologically based, are probably required for those patients who are destined for failure with the current approaches. Hence the future clinical research efforts should be directed toward developing reliable prognostic markers or techniques.

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