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Case Report

Primary Myxoid Liposarcoma of Breast with Round Cell Component - Report of A Rare Case with Review of Literature

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Abstract

Primary breast sarcomas are rare tumors constituting <1% of breast malignancies of which liposarcoma accounts for only 0.3%. A 58year-old female presented with a lump in the left breast of 2 years duration, gradually increasing in size and was associated with pain. On examination, she had a firm to hard large lump in the left breast involving all the quadrants. Fine Needle Aspiration Cytology was reported as phyllodes tumor. The patient underwent simple mastectomy. Grossly, the specimen was large, measuring 20.0×18.0×10.0 cm having a markedly variegated cut surface with solid and cystic areas. Solid areas appeared predominantly gelatinous. Histopathological examination of solid areas showed malignant spindle cell neoplasm with lipoblasts in myxoid stroma and numerous arborizing blood vessels. Focal areas had round cells with moderate amount of vacuolated cytoplasm. There were portions of necrosis and cystic degenerations. A diagnosis of primary myxoid liposarcoma was made and has been reported as it is a rare tumor in the breast with characteristic histopathology.

Keywords: Primary Breast Sarcoma; Myxoid Liposarcoma of Breast

Introduction

Malignant tumors of the breast predominantly comprise ductal carcinoma which is highly prevalent cancer in women worldwide. In comparison to carcinoma, primary sarcomas are very rare malignant tumors arising from the mesenchymal tissue of the breast, representing less than 1% of all breast malignancies [1]. Liposarcoma is an extremely rare tumor in the breast amounting to 0.3% of primary sarcomas arising from stromal tissue [2]. This malignant tumor arises from the interlobular stromal tissue of breast or can arise in an already existing cystosarcoma phyllodes [3]. The case reported here grew slowly to become large mass lesion in the breast and histopathologically had characteristic features of myxoid liposarcoma.

Case Report

A 58 year-old lady presented with a lump in the left breast of 2 years duration which was gradually progressive in size and was associated with pain. Examination revealed a firm to hard lump in the left breast measuring 20.0 × 15.0 cm involving all the quadrants (Figure 1a). However, axillary lymph nodes were not palpable. Fine Needle Aspiration Cytology (FNAC) was reported as spindle cell neoplasm and a possibility of Phyllodes tumor was suggested. Radiological findings also suggested the same and the patient underwent simple mastectomy.

Grossly, the specimen measured 23.0×19.0×11.0 cm, cut surface was variegated, grey white to yellow and tan with solid and cystic

areas along with soft necrotic portions (Figure 1b and 1c). One solid firm nodular portion showed gelatinous appearance (Figure 1d).

Routine processing of the multiple bits taken from various portions of the tumor including the surgical margins was done. Microscopy showed a malignant neoplasm comprising of lobules of spindle cells in a myxoid background. The tumour cells were spindle to satellite with many lipoblasts showing characteristic hyper chromatic nuclei indented due to vacuolations in the cytoplasm. The stroma showed numerous arborizing blood vessels giving chicken-wire appearance (Figure 2). Focal collections of round cells with high nucleo-cytoplasmic ratio and vacuolated cytoplasm were noted (Figure 3 a, b and e). There were a few bizarre cells with large pleomorphic nuclei and many abnormal mitotic figures were seen (Figure 3c). Cystic areas were surrounded by connective tissue lining with collections of haemosiderin laden macrophages and necrotic tissue debris in the cavity (Figure 3d). The surgical margins were free of tumor. Immunohistochemistry (IHC) showed strong positivity for S-100 which confirmed lipogenic origin of the tumor cells. The tumour was diagnosed as primary myxoid liposarcoma of left breast with round cell component based on the characteristic histopathological features.

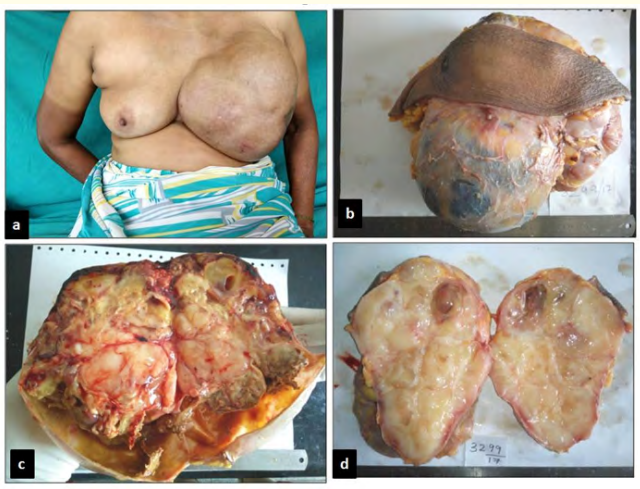

Figure 1: a) Large nodular mass in the left breast, b) Excised mass c) Variegated cut surface and d) Portion with gelatinous appearance in myxoid area.

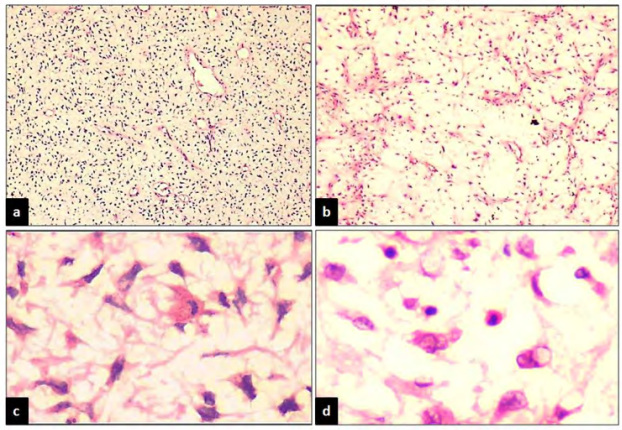

Figure 2: a) Spindle cells with myxoid tissue, b) Branching blood vessels with chicken wire pattern, (H & E, x100) c) Stellate cells and abnormal mitosis and d) Lipoblasts with cytoplasmic vacuoles compressing nucleus (H & E, x400).

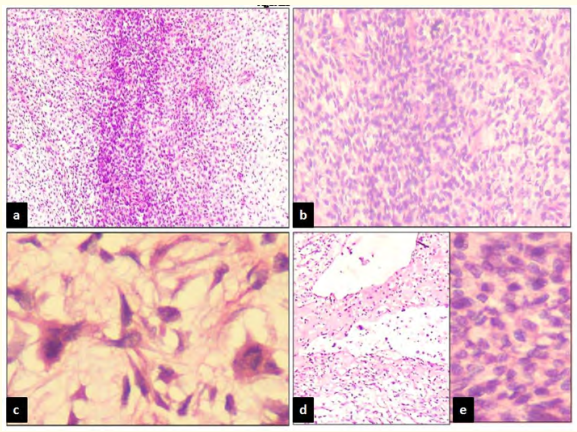

Figure 3: Myxoid liposarcoma a and b (H & E, x100) and e (H & E, x400) showing round cell component and c) Stellate cells in a myxoid stroma and d) Cystic areas (H & E, x100).

Discussion

Primary sarcomas arising from the mesenchyme of the breast are very rare tumors when compared to carcinomas of the breast which rank amongst the most prevalent types of cancer in women worldwide. Microscopically, most breast sarcomas comprise malignant fibrous histiocytomas followed by liposarcomas and fibrosarcomas. Liposarcoma which is the commonest type of soft tissue sarcoma is exceptionally rare in breast comprising just 0.0006% of

primary breast malignancies as per the review made by Adem., *et al.* and only 6.3% of breast sarcomas (19 of 304) were liposarcomas.[1] There are about 44 primary liposarcomas of breast reported in literature with well differentiated type as the commonest variant and the prominent differential diagnoses are malignant phyllodes tumor with liposarcomatous differentiation and high-grade metaplastic breast carcinoma [4]. Austin and Dupree reported the largest series with 13 cases of primary breast liposarcomas [5]. Primary liposarcomas of breast occur between 19 to 76 years with a median age of 47 years [6].

Most of the times Liposarcoma of the breast presents as unilateral mass lesion and involvement of axillary lymph nodes and the overlying skin are very rare. In the case presented here also there was no involvement of axillary lymphnodes. In comparison to the rapidly growing malignant phyllodes tumor, primary liposarcoma of breast is a slowly growing mass associated with pain as in the case presented here [7]. Grossly these tumors have a median size of 8.0 cm and may attain very large size becoming up to 20 cm in measurement [8]. Lipoblasts are neoplastic cells seen in liposarcomas having round shape, cytoplasmic lipid vacuoles with the nuclei typically indented showing scalloping effect by the vacuoles. These cells have to be differentiated from mimickers such as signet ring cells, hibernoma cells, necrotic and atrophic fat cells and also from the reactive fat cells or artifacts in cases of silicone implants of breast.

Austin., *et al*. have reported the largest case series of Primary liposarcoma of breast and reiterated that they are usually slow growing tumors which are painful, with no lymph node metastasis. They typically exhibit yellow-tan cut surface with gelatinous texture. On microscopy, lipoblasts have scalloped, irregular hyperchromatic nuclei with sharply defined intracytoplasmic vacuoles. The tumor also shows a characteristic chicken-wire branching of blood vessels. The important differential diagnoses of primary breast sarcomas are malignant phyllodes tumor (MPT) with heterologous liposarcomatous differentiation and high-grade metaplastic breast carcinoma (MBC).

The heterologous sarcomatous differentiation in MPT is most frequently liposarcomatous, although fibrosarcomatous, osteosarcomatous, rhabdomyosarcomatous and chondrosarcomatous differentiations have been described. Key histologic features in MPT that aid in the differential diagnosis are the characteristic leaflike fronds with stromal expansion and benign epithelial lining of phyllodes tumor. However, sarcomatous overgrowth and availability of limited tissue in core needle biopsy sampling may make identification of this architecture difficult [1].

High-grade MBC may be morphologically indistinguishable from dedifferentiated liposarcoma. MBC has poor prognosis and is the subtype constituting less than 1% of breast carcinomas. It includes a heterogeneous group of poorly differentiated carcinomas with spindle cells having squamous, chondroid and/or osseous elements. The spindle cell subtype of MBC is morphologically most similar to dedifferentiated liposarcoma.

MBC typically lacks true liposarcomatous differentiation but may display heterologous mesenchymal differentiation which can be morphologically indistinguishable from dedifferentiated liposarcoma. Histologic features that support the diagnosis of MBC are an associated epithelial components or ductal carcinoma *in situ* [1].

It is reported that Myxoid liposarcoma and round cell liposarcoma sharing genetic defect in the form of truncated *TLS* and *CHOP* genes. Pleomorphic variant showed MDM2 amplification in one-third of the patients. In Myxoid liposarcoma recurrent chromosome translocation t(12;16)(ql3;pll) has been widely reported as a specific marker [9].

Conclusion

Myxoid liposarcoma of breast has similar morphological features as its counterpart in other soft tissue locations and very rarely can have a round cell component. S-100 staining helps in ruling out non lipogenic tumors and this rare entity should be considered in the differential diagnosis of large painful breast tumours.

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