

CASE REPORT

Myxoid Liposarcoma: A Rare Soft Tissue Tumour in the Breast

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ABSTRACT

Liposarcoma is one of the most common mesenchymal tumour in adults but it is rare to occur in the breast. Our case was a 50 year old single nulliparous woman who presented with a right breast mass for one year duration. The mass was progressively increasing in size in the last few months. Breast examination showed a huge mass measuring 5 x 8 x 6 cm occupying the entire right breast. Mammogram showed a large homogenous soft tissue mass occupying the entire right breast with foci of calcification. A trucut biopsy showed a cellular tumour which was thought to be an invasive carcinoma. The patient underwent right modified radical mastectomy with axillary clearance. Macroscopy showed a well circumscribed lobulated solid haemorrhagic yellowish tumour mass measuring 180 x 110 x 50 mm. Microscopically the tumour was heterogenous comprising cellular round nonlipogenic mesenchymal cells and loose myxoid areas containing small cells. The typical arborizing 'chicken wire' capillaries were observed. Vacuolated lipoblasts were seen. All eleven axillary lymph nodes sampled showed no metastasis. A diagnosis of a myxoid liposarcoma was made. To raise the suspicion of a possible mesenchymal tumour, it is very important for clinicians to relay the clinical and radiological findings to the pathologist to avoid misdiagnosis in a trucut biopsy.

Keywords: Liposarcoma, Breast, Myxoid

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INTRODUCTION

Liposarcoma comprises less than 1% of all breast malignancies and represents 3-24% of the primary breast sarcomas (1). It may arise de novo as a primary liposarcoma or following radiation therapy. It may also occur as part of malignant phyllodes tumour (1). It usually presents as a slowly enlarging mass and is predominant in 5th to 7th decade of life. It appears as a well circumscribed dense mass on mammography. A well differentiated liposarcoma shows good prognosis if completely excised as compared to a pleomorphic liposarcoma which has a more aggressive behaviour.

CASE REPORT

A 50 year old woman, who was deaf and mute, but having normal mental state, presented to a local hospital with a huge breast mass of six months duration. The mass was progressively increasing in size. Examination

revealed a huge firm right breast mass measuring 18 x 11 cm. No skin or nipple changes were observed. Mammogram showed an opaque mass occupying the entire right breast. A trucut biopsy showed a cellular tumour comprising small round cells which was reported as invasive carcinoma. The patient underwent a right modified radical mastectomy with axillary clearance. Gross examination of the resected right breast revealed a well circumscribed lobulated solid yellowish tumour mass with haemorrhagic areas measuring 180 x 110 x 50 mm. A morphological diagnosis of a round cell myxoid liposarcoma was made. Eleven axillary lymph nodes sampled show no tumour involvement. All surgical margins and the nipple were tumour free, however the tumour mass measured 3 mm to the nearest surgical margin. The patient was referred to the oncologist and was given adjuvant radiotherapy to the chest wall after surgery, which she completed without any complications. No chemotherapy was given.

Pathology

Grossly the excised breast mass was composed of yellow to tan coloured well circumscribed tumour with haemorrhagic and necrotic areas (Figure 1).

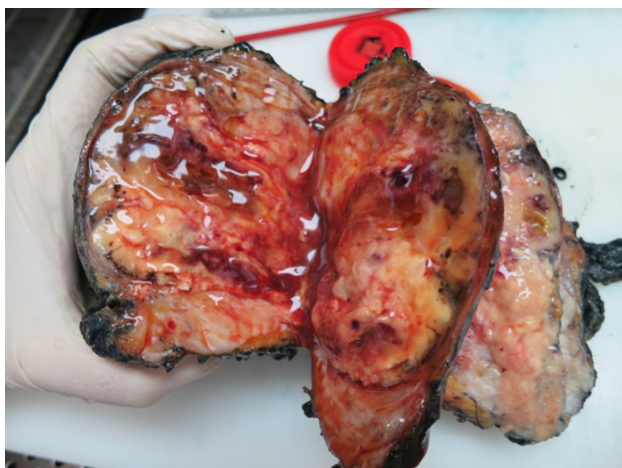


Figure 1 : Cross section of the mastectomy specimen showing a yellow to tan coloured well circumscribed tumour with haemorrhagic and necrotic areas.

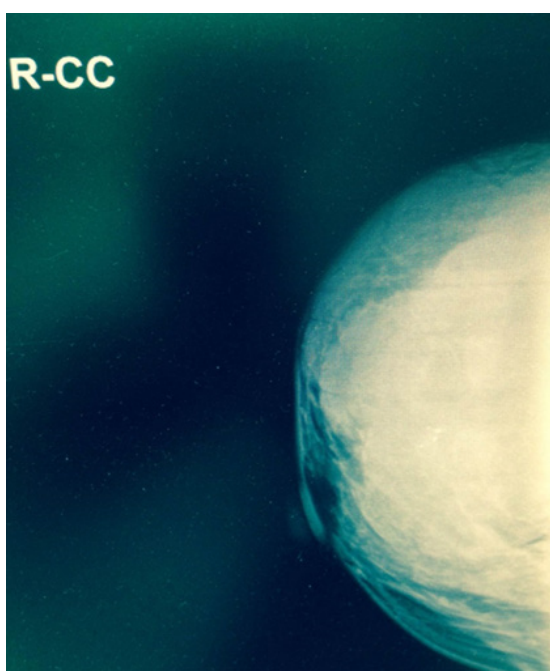


Figure 2 : Mammogram showing a large opaque mass occupying the entire right breast.

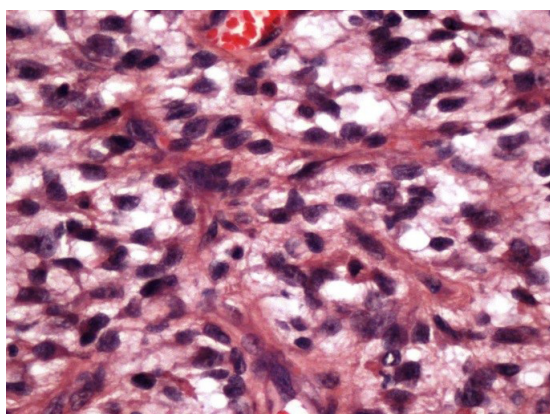


Figure 3 : Microscopically the tumour is composed of a heterogeneous population of cells comprising cellular round mildly pleomorphic nonlipogenic mesenchymal cells and a looser myxoid area containing small cells with surrounding arborizing ‘chicken wire’ capillary vasculature. Vacuolated lipoblasts are seen (H&E x 200).

Microscopy showed a heterogeneous population of cells comprising cellular round mesenchymal cells and a looser myxoid area containing small cells with surrounding arborizing capillary vasculature. Typical lipoblasts were seen (Figure 3).

DISCUSSION

Mesenchymal tumours of the breast are very rare as compared to epithelial tumours. Breast liposarcomas range from 3-20 cm in size with well circumscribed pushing borders. The cut surface is solid grayish white to yellow with areas of necrosis and cystic degeneration. A gelatinous cut surface suggests a myxoid component. Careful attention to clinical history as well as gross and microscopic details are important in making a correct diagnosis. The mammographic representation of a primary liposarcoma of the breast often shows a limited, round, oval or polycystic opacity, which may resemble a benign fibroadenoma. Mammographic findings which were very important findings were not made available to the pathologists when trucut biopsy was performed. This could be one of the factors which led to a wrong diagnosis of an invasive carcinoma in the small trucut biopsy sample. Biopsy may have hit a cellular tumour area which may look like an invasive carcinoma. In invasive carcinoma, the tumour is rather unlikely to occupy the entire breast as in this case, unless for a very aggressive and longstanding tumour. Even in a biopsy sample, if there is high clinical suspicion of a liposarcoma, S-100 immunohistochemistry could be performed to differentiate it from a carcinoma. In this case, S-100 immunohistochemistry (IHC) was performed which stained the lipoblasts and the intervening fat cells which acted as internal positive controls. MDM2 IHC which is characteristic for well differentiated and dedifferentiated liposarcomas was not performed. Next generation sequencing (NGS) was also not performed due to its high cost.

Liposarcoma is classified into four major histological subtypes: well-differentiated, myxoid/round cell, dedifferentiated, and pleomorphic (2). Well differentiated liposarcomas show variably sized adipocytes with fibrous septae transecting the tumour. The fibrous septae and adipose tissue components contain scattered atypical cells with hyperchromatic nuclei which are known as lipoblasts. Lipoblasts are highly characteristic but not specific for liposarcoma. Well differentiated liposarcoma has good prognosis and no distant metastasis.

Dedifferentiated liposarcoma is when a well-differentiated liposarcoma shows abrupt transition of its lipogenic component into a non-lipogenic component resembling a high-grade sarcoma (3). Myxoid liposarcoma shows proliferating small round-to-oval monotonous mesenchymal cells in a background of myxoid matrix with thin, arborizing

(chicken-wire or crow's feet) vessels. Mitoses and scattered lipoblasts can be seen. Myxoid liposarcoma can be mistaken for myxofibrosarcoma or extraskeletal myxoid chondrosarcoma (4).

Differential diagnoses of a liposarcoma include conditions which show presence of vacuolated cell resembling lipoblasts ie. fat necrosis, silicon granuloma, signet ring carcinoma and myxoid malignant fibrous histiocytoma. Typical lipoblasts can be recognised by their scalloped, irregular and hyperchromatic nuclei and well defined intracytoplasmic vacuoles. Typical lipoblasts and occasional positivity for S-100 protein in the spindle or poorly differentiated areas are the key feature in differentiating liposarcoma from non lipogenic tumours. Epithelial elements should not be seen in liposarcomas. Presence of epithelial elements raises the possibility of a metaplastic carcinoma or a phyllodes tumour with liposarcomatous differentiation. Therefore a careful search for epithelial elements such as glandular elements should be made to exclude metaplastic carcinoma.

Other than S-100 proteins, liposarcomas are also positive for p16 and CDK4⁵. Molecular studies show translocation t(12;16)(q13;p11) (fusion of CHOP (DDIT3) and FUS gene is a specific marker for myxoid liposarcoma. A history of breast radiation for any tumour type also does point towards the possibility of postradiation sarcoma, especially angiosarcoma. Other postradiation sarcomas include osteosarcoma, undifferentiated pleomorphic sarcoma (aka malignant fibrous histiocytoma), and fibrosarcoma (3). In this patient, there was no history of radiation to the breast or any other parts of the body.

Thus far, there is no treatment protocol that has been established for breast liposarcoma. There are no clear advantage of treating using either radical mastectomy, simple mastectomy or local excision. Removal of axillary tail is not necessary unless there are clear evidence that this procedure is needed for complete excision (5). For this patient, a right modified radical mastectomy was performed as well as an axillary lymph nodes sampling. A neoadjuvant radiotherapy has been shown to be effective in preventing local recurrence (5). In this patient, neoadjuvant radiotherapy was not performed. Chemotherapy was offered by the oncologist however due to the patient's inability to communicate (deaf and mute), chemotherapy was not favoured by carers, hence was not performed. A follow-up scheduled one year later showed no signs of recurrence or metastasis. Due to the low number of cases reported, prognosis for liposarcoma of the breast is not easily predictable.

CONCLUSION

The clinical and radiological findings may be useful in raising the suspicion of a sarcoma. Communication between clinicians and pathologists is important to prevent the likelihood of a wrong diagnosis on a biopsy. Distinguishing subtypes of primary breast sarcoma is relevant as certain subtypes may have poorer prognosis.

ACKNOWLEDGEMENT

We thank Quantum Diagnostics Sdn Bhd for their permission to publish this case. We also thank Columbia Asia Hospital, Seremban for providing us this case to be reported as a rare disease to occur in the breast. The abstract of this case report has been presented as a proceeding at the International Congress of Pathology and Laboratory Medicine, Shangri La Hotel, Kuala Lumpur, 26-28th August 2014.

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