CASE REPORT

Angiosarcoma of the Male Breast: A Case Report with Review of Literature

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Abstract
Primary breast angiosarcoma is a rare type of breast malignancy accounting for only 0.04% of all malignant breast tumors. It usually occurs in third to fourth decade and reported mainly in women and rarely in men. The histological features of angiosarcomas of the breast are conventionally grade I, II or III. Total mastectomy appears to be the main stay of treatment conferring benefit, chemotherapy and radiation therapy being of little proven value to date. The prognosis remains poor irrespective of grade of tumor.

Key Words
Angiosarcoma, Vascular Channels, Spindle Cells

Introduction
Malignant breast lesions arising from stromal tissues are extremely rare, accounting for <1% of all malignant breast tumors. Angiosarcoma is the most common sarcoma of the breast accounting for 0.04% of all malignant breast tumors (1). Its incidence among breast sarcomas varies from 2.7% to 9.1% (2). It usually presents as a painless, non tender palpable mass, with or without bluish discolouration of the skin. This malignant tumor occurs primarily in young women between 20 to 50 years of age group (3). Angiosarcoma occurs almost exclusively in female breast with rare cases of male breast angiosarcoma reported till date.

Preoperative diagnosis of angiosarcoma of the breast by aspiration cytology is often difficult (4). Aspiration smears are hemorrhagic and vary in cellularity. Well differentiated angiosarcomas tend to yield less material on aspiration, while intermediate and high grade tend to be more cellular (5). On cytology, angiosarcoma shows spindle, round to oval, epithelioid, pleomorphic cells.

This neoplasm carries a very poor prognosis. Metastases have been reported in the liver, lungs, skin, bones, central nervous system, spleen, ovaries and lymph nodes (6,7). This case is being reported for its rare histology and rare presentation in the male breast.

Case Report
A 24 years old male presented in surgical OPD of ASCOMS hospital, with a rapidly progressing lump in the left breast, lower outer quadrant, over a period of one month. There was a history of surgery at the same site 2 years back. There were no old records of subsequent histology with the patient. On examination, there was 2cm x 1cm mass in the lower outer quadrant of the left breast, mobile, firm and non tender. There was no evidence of nipple retraction, or axillary lymph node enlargement. Complete blood counts and X-Ray chest were within normal limits. Sonography (Fig 1) of the breast lesion showed architectural distortion with few linear anechoic curvilinear structures showing intraluminal echogenic contents.

Fine needle aspiration cytology was inconclusive as it yielded only blood and few oval to spindle cells. Subsequently incisional biopsy was done. Histological examination by study of haematoxylin and eosin sections showed vascular lesion composed of intercommunicating and anastomosing vascular channels lined by atypical endothelial cells (Fig2).

The stroma showed few haemosiderin laden macrophages, spindle cells and fibrous tissue. However, no mitotic figures were identified. There were multiple foci of fibrinoid material possibly due to previous FNAC procedures and the case was diagnosed as low grade Angiosarcoma (Grade 1).

Thereafter, patient underwent total mastectomy. The lesional area was again subjected to histopathological
examination and the same diagnosis was given. However, no lymph node involvement was there. The patient is currently undergoing chemotherapy.

Discussion

Angiosarcoma accounts for 0.04% of primary breast tumors and approximately 8% of breast sarcomas. Angiosarcoma is the most common sarcoma of the breast occurring almost exclusively in female breast and is extremely rare in male breast. Our case is a rare one because of the presence in a male patient. Sondenae et al reported angiosarcoma in female breast.

Melhouf et al reported angiosarcoma in patients 30-40 years of age. However, our patient was comparatively young -24 years. Angiosarcoma may have insidious onset, presenting as a painless often discrete palpable mass that grows rapidly. Approximately 2% of patients may present with diffuse enlargement of breast. Nipple retraction, discharge or axillary node enlargements are generally absent contrary to commonly seen in invasive carcinoma of breast. In our case too, these clinical findings were absent. The right breast is more commonly involved than left breast. However, in our case the lesional breast was left. Preoperative diagnosis, by aspiration cytology may be difficult as was in our case. Chen et al reported that false negative rate of biopsy was 37%.

The histological features of angiosarcoma of the breast are classified into low, intermediate and high grade lesions. Angiosarcoma have a wide range of histological appearance ranging from well differentiated low grade tumor consisting of infiltrating bland vascular channels to poorly differentiated grade 3 tumors composed of pleomorphic spindle cells with necrosis and numerous mitotic figures with lakes of blood. The histological appearance of our case was well differentiated grade 1 tumour. Sondenae et al surveyed the metastatic site of previously reported cases and found that liver was the most common site followed by lungs, lymph nodes and bones (9,10,12). Fortunately, in our case no metastatic lesion was present at the time of surgery. Mastectomy and chemotherapy are the mainstay of treatment because of inherent diffuse infiltrative nature of diseases without the need for primary axillary dissection due to rare lymphatic spread (11). In our case, total mastectomy was done and patient is currently undergoing chemotherapy.

Conclusion

The importance of this index case is that angiosarcomas are extremely rare in male breast. There presentation in the left breast is rare as compared to the right breast.

References


**Fig 1. Sonography of Left breast Angiosarcoma**

**Fig 2. Low Grade Angiosarcoma**