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Case Report

Primary angiosarcoma of the breast: a rare entity

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ABSTRACT

Primary angiosarcoma of the breast is a rare form of breast cancer, accounting for only 0.04% of all malignant breast tumours. We report a 35-year-old woman who presented with a recurrent lump in the left breast for four months. Fine needle aspiration cytology (FNAC) was suspicious of angiosarcoma and this diagnosis was confirmed on core needle biopsy. Modified radical mastectomy was performed as sole treatment. No chemo radiotherapy was given as tumor was grade I, margins were negative and there was no lymph node involvement. She is on regular follow up and no recurrence reported since the surgery. The 5-year disease-free survival for grade I tumours can be as high as 76%, and up to 70% for grade II tumours whereas for grade III tumours it is reportedly about 15%. Primary angiosarcoma of the breast is a rare malignancy with a poor prognosis, even after complete resection. Surgery is the mainstay of treatment with a limited role for chemotherapy and radiotherapy.

Keywords: Angiosarcoma, Primary angiosarcoma of the breast

INTRODUCTION

Angiosarcomas are rare tumors of vascular origin. Sarcomas are extremely rare tumours, accounting for <1% of all malignant breast tumours. Incidence of Primary angiosarcoma of the breast has been found to be 0.05%. The first case of Primary angiosarcoma of the breast reported by Schmidt in 1887 and since then about 219 cases have been reported in literature.[1,2] They have a high mortality rate with only 10-20% of the patients surviving beyond 5 years. Secondary angiosarcomas of the breast are more common and occur after breast irradiation. Here we discuss a patient with primary angiosarcoma of the left breast.

CASE REPORT

35 year old lady, P1L1, with no known medical co-morbidities, came with a recurrent left breast lump of four month duration, occasionally associated with pain. She also noticed a reddish discoloration of the skin over the lump. She gave no history of breast irradiation after the previous surgery or of taking any hormonal supplements. There was history of a previous surgery with the biopsy being reported as Haemangioma.

On examination there was a lump in the left breast of about 6X6 cm involving the upper outer and upper inner quadrant. The lump was smooth-surfaced, firm in consistency, non-tender and not attached to the skin or deep structures. Nipple was not retracted and there was no evidence of skin thickening. Bluish discoloration of the skin was present over the region of the lump on the left breast (Figure 1). There was no axillary lymphadenopathy. Right breast was normal on examination. Routine blood investigations and a chest X-ray were within normal limits. Ultrasonography of the breast showed a hypoechoic lesion in the left breast. CT

scan showed a tumor of the left breast with vascular enhancement. The fat planes between the tumor and chest walls were maintained. Core needle biopsy showed a low to intermediate grade vascular neoplasm. Fine needle aspiration cytology revealed features of a spindle cell neoplasm.

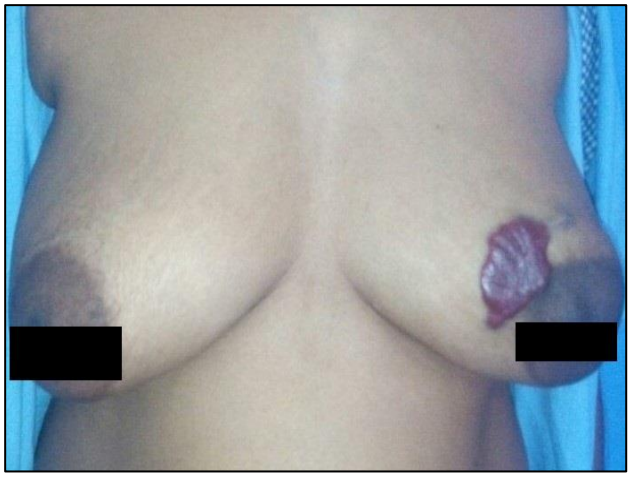

Figure 1: Clinical appearance of the lesion. a bluish discoloration of the skin above the area of the lump.

A left modified radical mastectomy was performed. Histopathological evaluation of the lesion showed a tumour comprising of anastomosing slit like vascular spaces with papillary projections at places infiltrating into the underlying breast parenchyma and fibroadipose tissue. These spaces were lined by cells exhibiting mild pleomorphism, oval nuclei and scant cytoplasm (Figure 2).

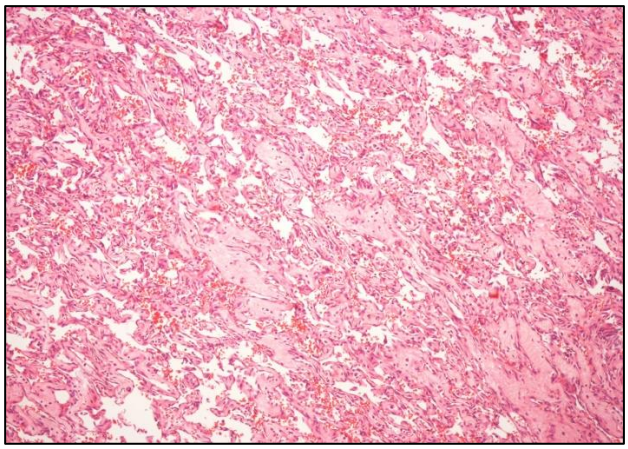

Figure 2: Vascular spaces with papillary projections at places infiltrating into the underlying breast parenchyma and fibroadipose tissue.

The Ki67 marker study revealed proliferative index of 60-70 %(Figure 4). CD 31 staining was positive (Figure 3) It was reported to be a well differentiated angiosarcoma (FNCLCC Grade 1) lymph nodes were negative for tumor, and tumor margins were free of tumor. Details were discussed with the hospital tumor board and she was not administered radiotherapy as the tumor was grade 1, margins were negative and there was no lymph node involvement. She is presently on regular follow up and there is no recurrence of the tumor.

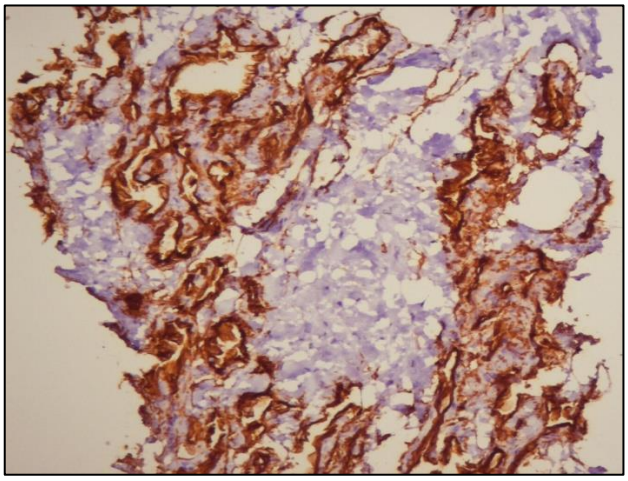

Figure 3: CD 31 immunostain, confirmatory for diagnosis of angiosarcoma.

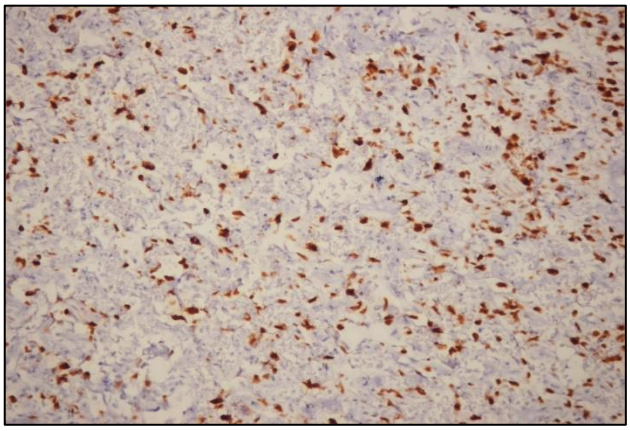

Figure 4: ki67 proliferative index- highly proliferative.

DISCUSSION

Primary angiosarcomas of the breast are a rare entity. Primary angiosarcomas usually occur in young women in the second to the fifth decade. Secondary angiosarcomas occurs in chronically lymphoedematous arms after axillary treatment for carcinoma (Stewart-Treves syndrome) and as a complication of breast radiation treatment.[3,4]

An uncommon clinical presentation of an angiosarcoma is spontaneous bleeding due to disseminated intravascular coagulation (DIC) by consumption coagulopathy, known as the Kasabach - Merritt syndrome. Primary angiosarcomas of the breast may present with a bluish discoloration of the skin with or without any lump.

Previous studies have found the tumor size and duration of symptoms have no bearing on the prognosis.[5-8]

The histologic features of angiosarcoma of the breast are classified into grades I, II and III. Angiosarcomas have a wide range of histological appearances from well-differentiated grade I tumours consisting infiltrating bland vascular channels to poorly differentiated grade III tumours with a sarcomatous spindle cell pattern. Large amounts of blood are often present. Papillary clusters of tumours cells, which can be mistaken for ductal carcinoma in situ, may also occur. Factor VIII and CD31 immunostaining confirm the diagnosis.[9]

Ultrasonography generally shows a solid mass that may have well - defined or lobulated margins, with both hypoechoic and hyperechoic appearance. There is often no acoustic shadowing. A contrast-enhanced MRI of the breast generally shows an enhancing vascular mass.[10]

Paclitaxel has been shown to produce excellent response in a number of studies in patients with primary angiosarcoma of breast. New agents against angiogenesis, such as bevacizumab or rapamycin, might also be useful against this tumour. Immunohistochemical staining for endoglin, known to be expressed mainly on the surface of endothelial cells, suggests the possibility of treating angiosarcoma with anti-endoglin monoclonal antibodies.[11]

Radiation treatment has been used in the adjuvant setting of breast sarcoma with the intent of improving both locoregional controls after surgical excision and survival. Even after mastectomy, radiotherapy has been thought to be beneficial for patient with microscopically positive margins.[12]

The prognosis, however, remains poor despite early diagnosis and treatment. The overall 5 year survival ranges from 8-50%. The 5 years disease free survival rate for low grade tumours can be as high as 76% and up to 70% for intermediate grade tumours. Whereas 5 years survival rate for high grade tumours is about 15%.[13]

CONCLUSION

Angiosarcoma of the breast though rare can be easily missed and the patient subjected to surgery without adequate investigation. FNA and biopsy remain gold standard for final diagnosis. A high index of suspicion and prompt intervention is hence necessary for better patient management and survival.

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