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Primary Angiosarcoma Breast - A Rare Case Report

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Abstract : Angiosarcoma, a rare malignant vascular tumor, accounts for less than 0.04 percent of all malignant lesions in breast. The incidence of primary breast angiosarcoma is about 17 new cases per million women. Previously, all angiosarcomas were considered to be highly aggressive but histologic stratification reveals quite different prognostic groups. The prognosis is usually poor because of the high rates of local recurrence and early development of metastases. Aggressive surgical resection is the mainstay of treatment. The role of adjuvant therapy has not yet been well established. This is a case report of Low grade Angiosarcoma of breast, which has a better survival of 91 for 5 years, according to WHO criteria.

Keyword : Primary Angiosarcoma Breast, Histological Grading, Prognosis.

INTRODUCTION: Primary breast sarcomas and angiosarcomas represent <1% and 0.04%, respectively of all breast cancers. Angiosarcomas arise de novo primary or secondary to radiotherapy treatments for breast cancer or postoperative lymphedema subsequent to a modified radical mastectomy. Only about 20% of angiosarcomas are primary sarcomas. They typically occur in younger women and arise from breast parenchyma. Secondary angiosarcomas usually occur in elderly women, arise from skin and show a pattern of infiltration into breast parenchyma from skin and subcutis. Both primary and secondary breast angiosarcomas carry a prognosis worse than mammary carcinoma.

CASE REPORT:

Clinical history:

A 40 year old female presented with painless swelling in right breast of 6 months duration which was gradually increasing in size and attained the present size. No history of pain or nipple discharge was noted. No history of previous radiotherapy was present. No family history of breast cancer was elicited.There was no relevent past history. On physical examination, a single swelling size of 8 x 6 cm in the upper outer quadrant of right breast was noted. It was not tender and not fixed to underlying structures. Skin surface was unremarkable.

FNAC REPORT: Few clusters of spindle cells showing mild pleomorphism in a bloody background noted. The possibility of Phyllodes tumor / Vascular lesion was given as initial report and excision biopsy was awaited

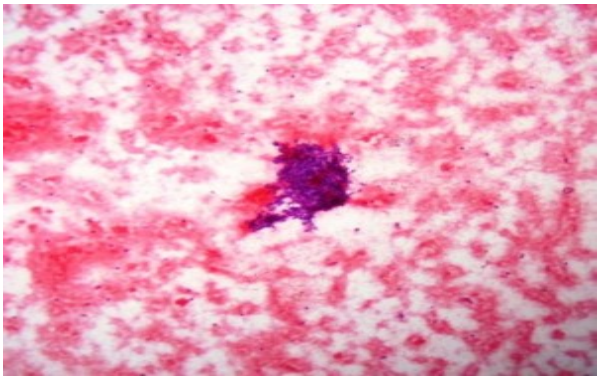

Fig.1 FNAC Low power view Cluster of Spindle cells in a bloody background

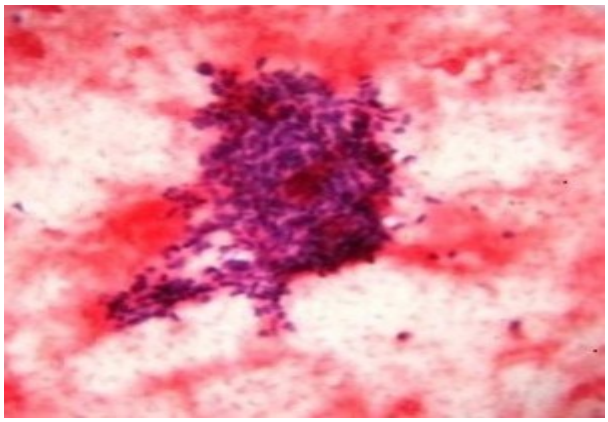

Fig.2 FNAC High Power view Cluster of Pleomorphic spindle cells

GROSS APPEARANCE: Wide local excision specimen of breast measuring 12 x 9 x 4 cms was received.

Cut surface: Cut surface of received breast specimen showed a irregular grey brown hemorrhagic tumor measuring 3.5 x1.3 x 2 cm. Tumor was spongy, grey brown with irregular tiny cystic spaces filled with hemorrhagic material infiltrating into adjacent fatty tissue.

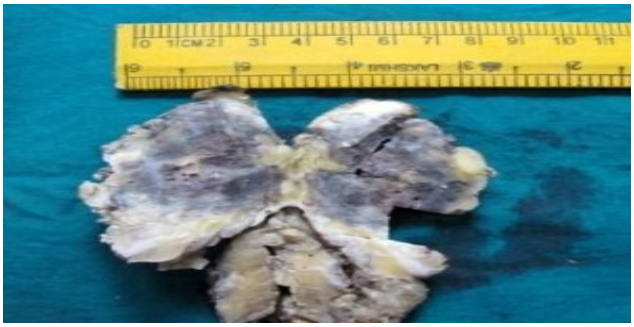

Fig.3 Gross Picture Irregular, spongy, grey brown tumor with tiny cystic spaces filled with haemorrhagic material

MICROSCOPY: Multiple sections studied from the tumor showed anastomosing open vascular channels with diffuse infiltration around and into lobular stroma and fat tissue. The vascular channels were lined by a single layer of flat endothelial cells, with hyperchromatic nucleus and increased nuclear cytoplasmic ratio. Occasional endothelial tufting was seen. No papillary form or solid nests were seen. Mitotic figures were rarely seen. No necrosis or blood lakes were present.

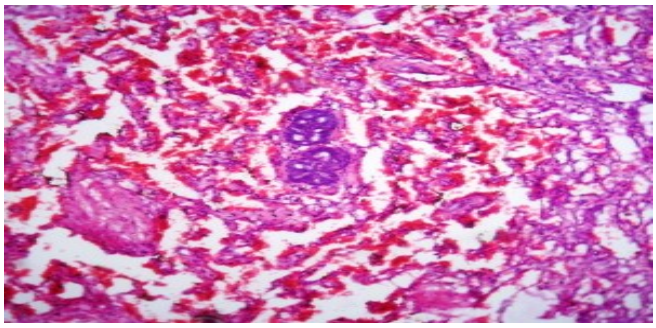

Fig.4 HPE low power view Anastamosing vascular channels filled with blood infiltrating breast parenchyma

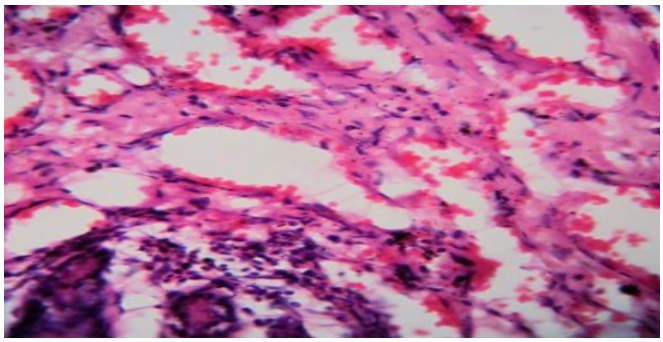

Fig. 5 HPE High power view Vascular spaces lined by hyper-chromatic and pleomorphic endothelial cells

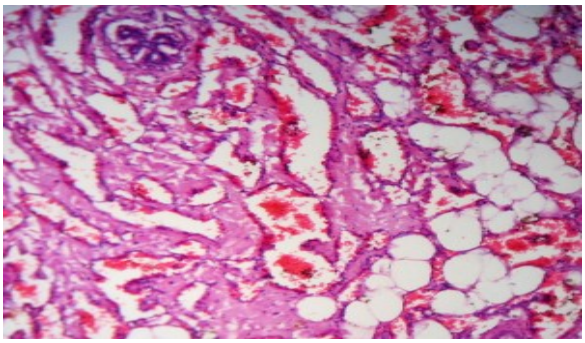

Fig.6 HPE Tumour infiltration into the surrounding adipose tissue

DIAGNOSIS : Low grade Angiosarcoma breast. All cut margins were free of tumor.

IMMUNOHISTOCHEMISTRY REPORT:

CD 31 - Membranous positivity of endothelial cells
CD 34 - Membranous positivity of endothelial cells

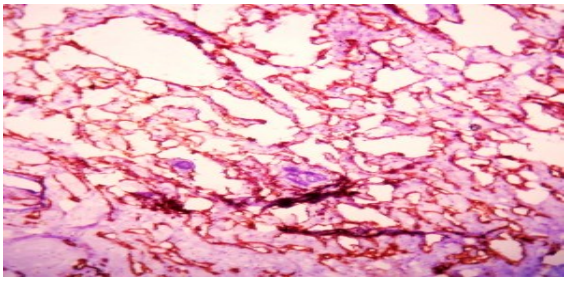

Fig 7. CD 31 Cytoplasmic positivity of endothelial cells

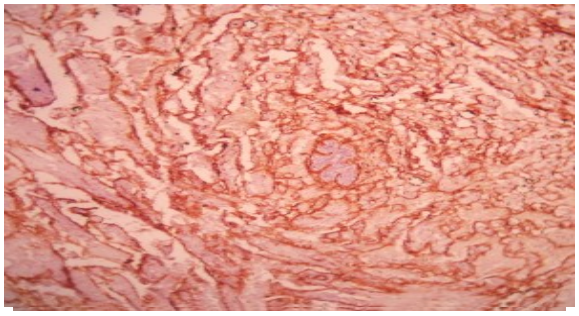

Fig.8 CD 34 Cytoplasmic positivity of endothelial cells

DISCUSSION: Breast angiosarcoma is observed as a primary neoplasm or secondary to breast-conserving surgery combined to radiation therapy. Secondary angiosarcomas of breast are more common than primary. Primary Breast angiosarcoma is more frequent in young women. Between 6 and 12% of primary breast angiosarcomas are diagnosed during or shortly after pregnancy. This suggests hormonal involvement. Complete surgical excision of breast angiosarcoma is the best course of treatment. Total mastectomy is the treatment of choice. Since hematogenous dissemination is more common, axillary node dissection is not indicated.

GRADING: Prognosis of the patients depends upon tumor grade, tumor size at diagnosis, and margin status at surgery. Histologically, Primary breast angiosarcomas are a heterogeneous group of lesions. They are composed of interanastomosing vascular channels lined by hyperchromatic endothelial cells, occurring within the breast parenchyma. Angiosarcomas of the breast can be classified into three main growth patterns, well-differentiated (Grade I), moderately differentiated (Grade II) and poorly differentiated (Grade III) tumors. The pattern of the low grade angiosarcoma resembles endothelial lining histologically. Hyperchromasia and nuclear pleomorphism are noted. In majority of high grade angisarcomas, endothelial tufting and papillary formations are prominent. The endothelial cells show marked cytological atypia with prominent nucleoli and frequent mitotic figures. Haemorrhage into the surrounding stroma resembling "blood lakes" and areas of necrosis may be present. However, variations in growth pattern are commonly found within a single lesion. High-grade lesions can have lower-grade areas at their peripheral and superficial aspects.

Donnell et al Grading system of Angiosarcomas

S.no		Low grade	Intermediate grade	High grade
1	Anastomosing vascular channels	Present	Present	Present
2	Hyperchromatic endothelial cell	Present	Present	Present
3	Endothelial tufting	Minimal	Present	prominent
4	Papillary formations	Absent	Focally present	Present
5	Solid & spindle cell foci	Absent	-/minimal	Present
6	Mitoses	Rare	Present in papillary areas	Numerous
7	Blood lakes	Absent	Absent	Present
8	Necrosis	Absent	Absent	Present

METASTASIS: The most common sites of metastasis of high grade angiosarcomas are lung, liver, bone, contralateral breast and skin.

PROGNOSIS AND PREDICTIVE FACTORS: Prognosis depends upon pathological grading. Patients with a grade I/II tumor are alive after 15 years. Grade II/III tumors are the most aggressive, and the median disease-free survival is about 15 months. Even though low grade angiosarcoma has a relatively favourable prognosis, there is a high rate of local recurrence and early development of metastases.

DIFFERENTIAL DIAGNOSIS: The main differential diagnoses of high-grade angiosarcoma are metaplastic carcinoma with angiosarcomatous differentiation, pseudoangiomatous hyperplasia breast, papillary endothelial hyperplasia and other poorly differentiated sarcomas. The use of immunohistochemical markers for endothelium, CD31, CD34 and factor VIII confirm most angiosarcomas including poorly differentiated ones. CD31 is the most specific marker for endothelial differentiation, whereas CD34 is more sensitive. The prognostic factors of breast angiosarcoma include tumor size, presence of residual disease, cellular pleomorphism and proliferative index. Aggressive surgical resection is the mainstay of treatment. The role of adjuvant therapy with radiotherapy and chemotherapy has not yet been well established.

CONCLUSION: Angiosarcomas are rare tumors of the breast; and primary angiosarcomas are still rarer. This casehad the typical features of Angiosarcoma of low grade in Histopathology and it was confirmed with Immunohistochemistry. Thorough sampling, early and precise histological grading and diagnosis are important. This caseshows the vital role of precise pathological diagnosis in guiding the clinical management.Even though these tumors have a bad prognosis, surgical treatment by total mastectomy is preferred.The aggressive nature of this disease demands careful follow-up of these patients. Due to rarity of thecondition the appropriate strategies of treatment remain to be determined.

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