



## A MASQUERADER IN THE BREAST-CASE REPORT

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**ABSTRACT** Primary Angiosarcoma of breast is a rare stromal sarcoma in the breast (1). We present this case for its rarity. The prognosis is poor because of the high rate of local recurrence and early development of metastasis. Total Mastectomy is recommended primary therapy with regular follow up.

We report a 43 year old female with left breast mass of 3 months duration. Mammogram showed fibrocystic change, MRI showed infiltrating malignancy. Histopathology of mastectomy specimen showed anastomosing vascular channels lined by pleomorphic spindle cells with moderate nuclear atypia and hyperchromasia. The tumour was dissecting through the breast parenchyma and fat. CD 34 and CD 31 were positive and ki 67 labelling index showed 20% of tumour cells positive. The diagnosis of low grade angiosarcoma was made.

**KEYWORDS :** Breast, Primary angiosarcoma, Histopathology, IHC

**INTRODUCTION-**

Primary angiosarcoma of breast is rare and accounts for <0.04% of all malignant neoplasms of the breast (2,10). Only about 20% of angiosarcomas are primary sarcomas. The incidence of primary breast angiosarcoma is 17 new cases per million women (1).

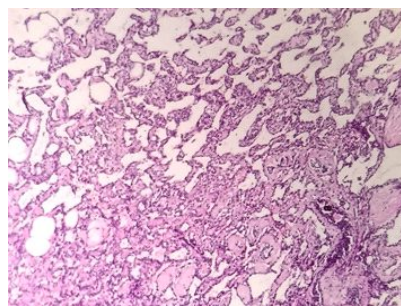
**CASE DESCRIPTION**

43 Year old female with left breast lump for 3 months, on clinical evaluation mass of size 10x8 cm was seen in the upper medial quadrant. Overlying skin was normal. No axillary lymphadenopathy. Contralateral breast was normal clinically. USG and Mammogram revealed fibrocystic disease. MRI suggestive of infiltrating malignancy, hence proceeded with FNAC which showed hemorrhagic aspirate material only. Then trucut biopsy of left breast was done and histopathological examination of 2 cores showed low grade angiosarcomatous features.

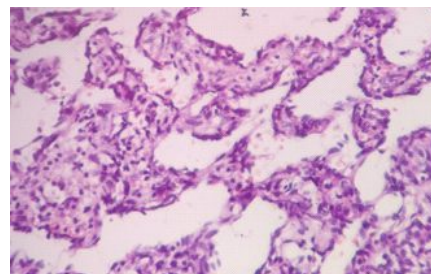
So patient was proceeded with left modified radical mastectomy, mastectomy specimen on (fresh state) measuring 26x21x7 cm on cutting the specimen around 300 ml of blood was drained. Cut surface showed a ill defined fleshy mass with many hemorrhagic spots with brownish areas in the upper medial quadrant, inking was done. After overnight fixation in 10% neutral buffered formalin, fleshy growth (9x 6.5x 6.5 cm) which was firm grey white with hemorrhagic spots and brownish areas. All margins from growth were noted and bits taken. 18 nodes made out. Histopathological examination showed a neoplasm composed of anastomosing vascular channels lined by pleomorphic spindle shaped cells exhibiting moderate nuclear atypia with hyperchromatic nuclei. Stroma shows lymphoplasmacytic infiltrate with areas of haemorrhage. The tumour cells dissect through the breast parenchyma. IHC showed positivity for CD 31 & CD 34 and ki 67 index showed 20% of tumour cells are positive.



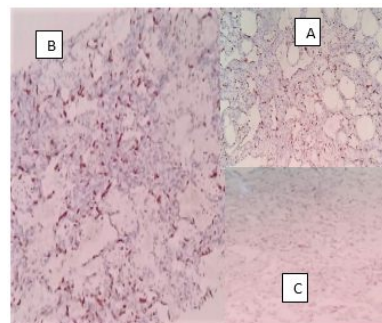
Gross photograph of mastectomy specimen showing hemorrhagic areas



HPE showing anastomosing vascular channels lined by pleomorphic endothelial cells with stromal lymphoplasmacytic infiltrate



High power view showing moderate nuclear atypia with hyperchromasia



A-CD 31 POSITIVE  
B-CD 34 POSITIVE  
C-KI 67 -20% POSITIVE

## DISCUSSION-

Primary angiosarcoma of the breast is extremely rare primary tumor arising in patient having no history of treatment for breast cancer, while secondary angiosarcoma occurs mostly after treatment with radiotherapy. Primary angiosarcoma affects relatively younger patients usually at the age of 20-50 years (6), with 6–12% of the cases occurs during pregnancy (3,5)

The **differential diagnosis** of Angiosarcoma breast include (1,2 ), benign neoplasms like hemangioma of breast, hemangiopericytoma, PASH, fibromatosis, myoepithelioma

Malignant tumours like, malignant phylloides, metaplastic carcinoma, squamous cell carcinoma with sarcomatoid features,

Hemangiopericytoma grossly has a well-circumscribed round-to-oval firm to hard mass with homogeneous pale yellow, gray, or white tissue. Cut surface has a whorled texture with dilated vascular spaces and nodular contour. Microscopically storiform pattern of arrangement of tumour cells, staghorn vessels and anastomosing vascular channels seen.

Hemangioma-size < 2 cm, separated into lobules by fibrous septae, usually has unconnected vascular channels very rarely has simple anastomosing vascular channels without atypia.

PASH- Grossly has homogenous fibrous and tan grey white areas. microscopically -empty anastomosing spaces with dense collagenised stroma. Myofibroblasts lining pseudoangiomatous spaces shows CD 34 reactivity.

Myoepithelioma-grossly nodular, microscopically dilated ducts with adenomatous papilloma. positive for calponin. Malignant phylloides-circumscribed grey white cauliflower like mass, rubbery with necrosis and haemorrhage .microscopically hypercellular stroma, stromal overgrowth with glandular elements and rarely contains angiosarcomatous elements. cytokeratin positive metaplastic carcinoma- more than one stromal elements will be seen (osseous, chondromyxoid...) angiosarcoma usually present as a painless rapidly growing palpable mass (13). Few patients complain of pain and tenderness and 2% of patients may present with diffuse enlargement of the breast. Sometimes bluish red discoloration of the overlying skin may be noted (8). Nipple retraction, discharge, and axillary lymphadenopathy are generally absent. Bilateral tumors are rare but may be seen specially post menopausal women (1)

Grossly Angiosarcoma varies in size from 1 to 20 cm or more, averaging about 5 cm. It forms a friable, firm or spongy hemorrhagic tumor. Areas of cystic hemorrhagic necrosis are commonly evident in large, high-grade lesions. In our case also fleshy growth (9x6.5x 6.5 cm) which is firm grey white with hemorrhagic spots and brownish areas present.

The cytological and radiological findings are often non specific for diagnosis. In our case FNAC shows haemorrhagic aspirate material only. Chen et al. (3) reported that false negative rate of FNAB is 37%. Histopathology plays an important role in diagnosis and grading of tumour Angiosarcoma was the only histologic type of sarcoma associated with a significantly poor prognosis(5).

Pathologically, angiosarcomas are subdivided into three groups according to the classification proposed by Donnel et al [8]. Grade I (well differentiated) contains open anastomosing vascular channels that proliferate within dermis, subcutaneous tissue or breast tissue, Angiosarcoma dissect through the stroma, causing distortion but little destruction of the preexisting lobules and ducts. The endothelial cells are usually flat; the nuclei is hyperchromatic and contains small nucleoli. Solid and spindle cell foci, blood lakes, and necrosis are absent.

Intermediate-grade angiosarcoma differs from low-grade by containing additional cellular foci of papillary formations with solid and spindle cell proliferation. The greater part of the tumor, however, is still composed of low-grade histology. Slightly increased mitotic activity is observed. In Rosen's study, intermediate-grade angiosarcomas behave more like low-grade sarcomas.

In grade III endothelial tufting and papillary formations are prominent.

Conspicuous solid and spindle cell areas, mostly devoid of vascular formations, are present as well. Mitoses may be brisk, especially in more cellular areas. Areas of hemorrhage, known as "blood lakes," and necrosis are also seen single layer of endothelial cells lines these channels .Ki67 shows >45 % positivity

The endothelial cells show reactivity for several markers, including CD31, CD34 and von Willebrand factor (factorVIII). Among them, CD31 is considered the most sensitive and most specific endothelial cell marker

The most frequent sites of metastases are bone, the lungs, the liver, the contralateral breast, and the skin, other than local recurrences at the mastectomy site

Total mastectomy is the recommended primary surgical therapy. axillary node dissection not recommended. Chemotherapy is observed to be beneficial in high-grade lesions and in the metastatic setting.

## CONCLUSION-

We presented this case for its rarity in literature and diagnostic difficulty, because radiologically and cytologically it may be misleading most of the time, and also it is a great masquerader of carcinoma and other benign lesions. Histopathology along with immunohistochemistry Is only definitive means of establishing diagnosis of angiosarcoma.

## REFERENCES

- Bennani et al.: Primary angiosarcoma of the breast: a case report. *Diagnostic Pathology* 2013 8:66.
- Athar M, Singh S, Chaudhary AK, Sachan M, Khan L. Primary angiosarcoma of the breast in a seventy-year-old female: a rare case report. *J Res Hosp* 2016;1:28
- Chen KTK, Kirkegaard DD, Bocian JJ: Angiosarcoma of the breast. *Cancer* 1980, 46:368371.
- Wang XY, Jakowski J, Tawfik OW, Thomas PA, Fan F: Angiosarcoma of the breast: a clinicopathologic analysis of cases from the last 10 years. *Ann Diagn Pathol* 2009, 13(3):147–150.
- Rosen PP, Kimmel M, Ernberger D. Mammary angiosarcoma. The prognostic significance of tumor differentiation. *Cancer* 1988;62:2145-51.
- Scow JS, Reynolds CA, Degnim AC, Petersen IA, Jakub JW, Boughey JC: Primary and secondary angiosarcoma of the breast: the Mayo Clinic experience. *J Surg Oncol* 2010, 101(5):401–407.
- Desbiens C, Hogue JC, Levesque Y: Primary Breast Angiosarcoma: Avoiding a Common Trap, *Case Reports in Oncological Medicine*. 2011;5. Article ID 517047.
- Donnel RM, Rosen PP, Liberman PH, et al. Angiosarcoma and other vascular tumours of the breast. *Am J Surg Pathol*. 1981;5:629-42.
- Scow JS, Reynolds CA, Degnim AC, Peterson IA, Jakub JW, Bonghey JC. Primary angiosarcoma of the breast: the Mayo Clinic experience. *Journal of Surgical Oncology*. 2010;101(5):401-07.
- Vemulapalli S, Mita A, Alvarado Y, et al. The emerging role of mammalian target of rapamycin inhibitors in the treatment of sarcomas. *Targeted Oncology*. 2011;6(1):29-39.
- A. Vorburger, Y. Xing, K. K. Hunt et al., —Angiosarcoma of the breast, *Cancer*, vol. 104, no.12, pp.2682–2688, 2005.
- R. C. Fields, R. L. Aft, W. E. Gillanders, T. J. Eberlein, and J. A. Margenthaler, — Treatment and outcomes of patients with primary breast sarcoma, *American Journal of Surgery*, vol. 196, no. 4, pp. 559–561, 2008.
- Johnson CM, Garguilo GA. Angiosarcoma of the breast: A case report and literature review. *Curr Surg* 2002;59:490–4.