



CASE REPORT

## An interesting pathological diagnosis: Angiosarcoma of breast

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**Abstract:** Primary angiosarcoma of breast is an extremely unusual variant of breast malignancies and its incidence is about 0.05% of all primary breast malignancies. Herein, we are presenting an unusual case of an 84 year old female patient with primary angiosarcoma of breast, which was diagnosed by fine needle aspiration cytology (FNAC) and confirmed with histopathology and immunohistochemistry approaches. She was treated with modified radical mastectomy (MRM), followed by radiotherapy and was observed to be recovering well based on the last 10 months of her post-treatment follow-up visit.

**Keywords:** Breast angiosarcoma; Immunohistochemistry; CD 31

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### Introduction

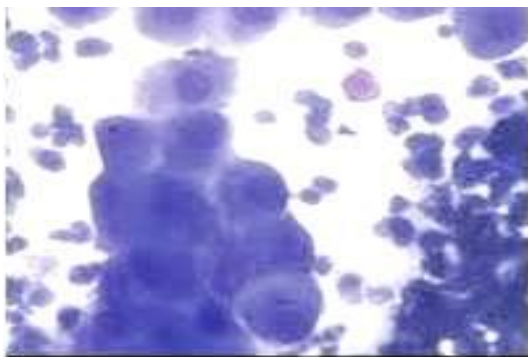
Primary mammary angiosarcomas arising from breast parenchyma are unusual and as per existing literature the incidence is 0.05% of all primary breast malignancies<sup>[1]</sup>. Breast carcinomas form the overwhelming majority while sarcomas are of very negligible incidence. The breast sarcomas include fibrosarcoma, malignant fibrous histiocytoma, angiosarcoma and liposarcoma<sup>[1-4]</sup>. Incidence of soft tissue sarcoma of breast is less than 1% of all breast cancers<sup>[3,4]</sup>. The incidence of post irradiation breast sarcoma is 0.01% to 0.02% per year and that of angiosarcoma of breast is 0.002% to 0.05% per year<sup>[4]</sup>.

### Core tip

Primary angiosarcoma of breast is an extremely unusual type of breast malignancy. Early clinical suspicion with adjuvant diagnostic tools (i.e. FNAC), and meticulous histopathological examination along with immunohistochemistry technique, is an effective diagnostic approach in order to have a better outcome for the patient.

### Case Report

An 84 year old female patient was admitted to Yashwantrao Chavan Rural Hospital, Latur with complaints of painless and rapidly growing lump in her right breast since two months prior. There was no history of previous surgery or radiotherapy and there was no history of exposure to vinylchloride, arsenic thorotrast or sexual hormones. Based on clinical examination, a non-tender lump measuring 10 cm wide and 8 cm long was seen fixed to the chest wall. The overlying skin, nipple and areola were normal and there was no evidence of either bluish or reddish discoloration. There were no palpable axillary lymph nodes. Ultrasonography (USG) revealed benign neoplastic lesion suggestive of haemangioma. FNAC was planned and performed. The microscopy revealed loosely cohesive malignant cells with hyperchromatic bizarre nuclei in the background of blood reported as malignant lesion (*Figure 1*). The complete body bone scintigraphy did not reveal any lesion suspected of metastasis elsewhere in the bones of the body. The whole body scan via positron emission

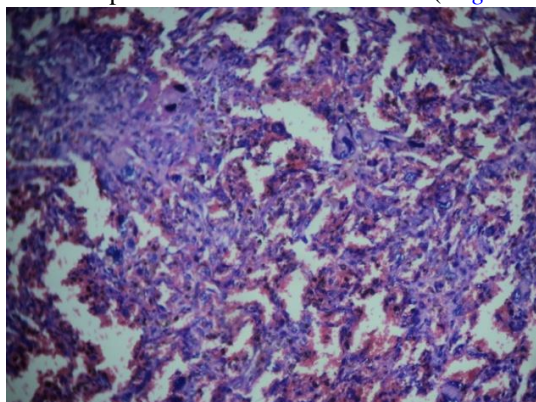


*Figure 1* Photomicrograph of dyscohesive malignant cells. Gross specimen showing multiple pieces of soft, friable, and spongy hemorrhagic mass (hemorrhagic with reddish brown discoloration)

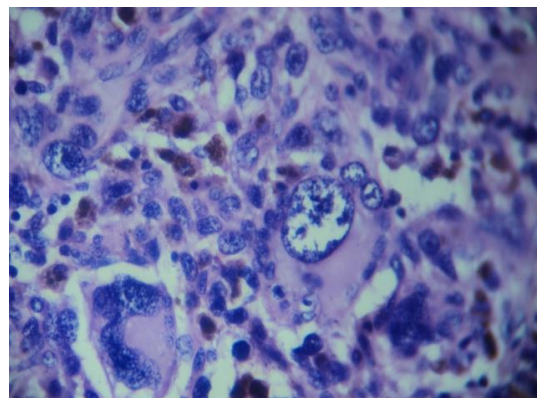
tomography (PET) did not reveal any secondary or suspected lesion.

In view of FNAC report, core biopsy was planned and performed for definitive histological diagnosis and classification of the tumour. Photomicrograph of the core biopsy revealed a tumour composed of anastomatic vascular channels lined by endothelial cells which showed prominent hyperchromatic pleomorphic nuclei exhibiting prominent nucleoli. Lumina of the neoplastic vessels were filled with red blood cells (RBCs). These vascular channels were intermingled with solid endothelial and spindle cell areas that showed necrotic foci and numerous mitotic figures. Multinucleated highly pleomorphic giant cells were also observed. The solid cellular component was more than 50% of total neoplastic area. Large areas of necrosis and haemorrhages were evident. Infiltration of the tumour into its adjacent muscle and fibrofatty tissue was also observed (*Figures 2 and 3*).

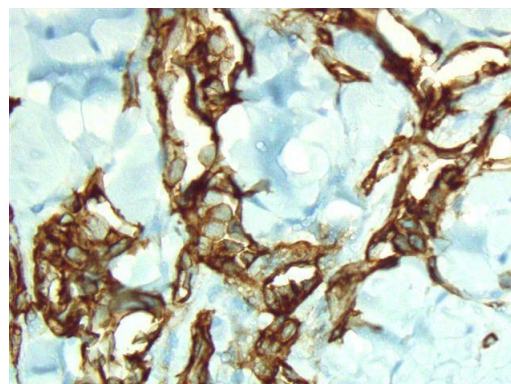
Immunohistochemical result showed CD31 immunopositive expression in the tumour (*Figure 4*).



*Figure 2* Low power view at 10X magnification showing inter-anastomosing vascular channels intermingled with solid endothelial and spindle cell areas



*Figure 3* High power view at 40X magnification showing highly pleomorphic giant cells



*Figure 4* Endothelial cells showing immunopositive expression for CD 31

Finally we arrived at the diagnosis of primary angiosarcoma of breast and treated accordingly with modified radical mastectomy followed by radiotherapy dose-fractionation of 60 Gy. The patient was diagnosed with breast cancer recurrence-free with no metastasis evident since the last 10 months of her post-treatment follow-up visit.

## Discussion

Angiosarcoma was formerly known as hemangiosarcoma, hemangioblastoma or lymphangiosarcoma. At present, however, there are no reliable criteria to make a histological distinction between tumours derived from endothelium of blood vessels and lymphatic vessels<sup>[1]</sup>. Mammary angiosarcoma can be subdivided into four categories:

1. Primary (*de novo*) angiosarcoma forms in breast parenchyma.
2. Secondary angiosarcoma in skin and soft tissue of arm following ipsilateral radical mastectomy and subsequent lymphoedema—the Stewart Treves (S-T) syndrome.

3. Secondary angiosarcoma in skin and chest wall following radical mastectomy and focal radiotherapy.

4. Secondary angiosarcoma in skin or breast parenchyma or both following conservative treatment and radiotherapy<sup>[1]</sup>.

The age of the patients with primary angiosarcoma ranges from 17–70 years old, with the median age being 38 years. The average age of patient with grade III angiosarcoma is 29 years old. In the present case, this tumour was identified in an 84 year old patient. Breast angiosarcoma usually presents as painless palpable fast growing mass<sup>[2]</sup>. Approximately 12% of patients possess diffuse breast involvement. Large tumors may be associated with ecchymosis like skin changes. In our case, the tumour was deeply located in the breast tissue with absence of skin involvement<sup>[2]</sup>. Angiosarcomas vary in size from 1–20 cm with the average size of 5 cm, besides retaining a sponge like appearance<sup>[4]</sup>. Poorly differentiated tumors are ill-defined, indurated fibrous lesion. Angiosarcomas of breast are graded as grade I (well differentiated), grade II (intermediately differentiated), and grade III (poorly differentiated). In grade III, > 50% is solid area with spindle cell differentiation without evidence of vascular channels<sup>[1]</sup>. The present case is of poorly differentiated (grade III) angiosarcoma.

The antibodies commonly used for endothelial cells are factor VIII, CD34 and CD31. CD31 seems to be the most sensitive and specific for endothelial differentiation. Endothelial marker study by immunostaining is must to arrive at final diagnosis<sup>[5,6]</sup>. The markers are positive usually in grade I and II. However, negative in poorly differentiated histological features. Our case showed strong immunopositive expression for CD31 via immunohistochemistry approach and the diagnosis was confirmed as primary angiosarcoma of breast. Tumours of both low- and intermediate-grade should be differentiated from haemangioma<sup>[1]</sup>.

Prognosis of well differentiated angiosarcoma (better, intermediate and high grade) is usually lethal<sup>[1]</sup>. Survival probabilities of being disease-free five years after initial treatment are estimated as follows: Grade I: 76%, Grade II: 70% and Grade III: 15%. The median length of disease-free survival is also related to tumour type i.e., Grade I >15 years, Grade II >12 years, and Grade III: 15 months<sup>[7]</sup>. Metastasis takes place mainly in lung, skin, contralateral breast, liver and bone. Axillary lymph node involvement is very rare. Seemingly, metastasis was not seen in our case.

In conclusion, primary angiosarcoma of breast is an

extremely unusual variant of breast malignancies. We are presenting this case in view of its unusual presentation and rare site of involvement. Meticulous core biopsy examination and immunohistochemistry approach is an effective diagnostic tool to differentiate primary angiosarcoma of breast from other malignant lesions of breast.

## Author Contributions

Sachin B Ingle wrote the first draft, critically revised the intellectual content and gave final approval of manuscript. DD Girji diagnosed the case based on histopathology.

## Conflict of interest

The authors declared no potential conflict of interest with respect to the research, authorship, and/or publication of this article.

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