ISSN: 2634 - 680X

Journal of Clinical Case Studies Reviews & Reports

SCIENTIFIC Research and Community

Case Report

Open Access

Primary Angiosarcoma of Breast – A Rare Case Presentation

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SUMMARY

Breast sarcomas are histologically heterogenous group of non- epithelial malignancies arising from mesenchymal component of the breast. Angiosarcoma of the breast constitutes 1 % of all soft tissue tumour of the breast and primary angiosarcoma is an extremely rare entity with an incidence of 0.05% of all the breast tumour. We report a case of primary angiosarcoma of the breast in a 57 year old woman, with no previous radiotherapy and is treated with breast conservative surgery.

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Received: March 20, 2021; **Accepted:** March 27, 2021; **Published:** March 30, 2021

Keywords: Angiosarcoma, Radiotherapy, Breast Conservative Surgery

Introduction

Primary Angiosarcoma of the breast is a rare neoplasm with an incidence of <0.05% of all malignant breast tumour. Angiosarcoma arises de novo with a median age at onset of 40 years or can be secondary to radiation treatment of an epithelial breast cancer with a median onset age of 70 years (median of 10.5 years after radiotherapy)[1].

It carries poor prognosis and no specific radiological features are diagnostic to it. They are highly malignant, grows at faster rate and evolve to rapid local recurrence and appearance of visceral metastasis. The treatment of mammary sarcomas is simple mastectomy or wide local excision of the lump.

There are 3 main histopathological patterns of Angiosarcoma: type I, characterized by vascular channels invading the breast tissue with scarce endothelial proliferation; type II, presenting patterns of papillary endothelial components and type III, with evidence of endothelial components, necrosis and haemorrhage [2].

Available literature pertaining to this case are predominantly post radiation, and very few have been reported as primary angiosarcoma.

Here we report a case in a 57 year old female presenting with breast lump since one month and on the basis of histologically and immunohistochemical examination, it is diagnosed as primary angiosarcoma of the breast.

Case History

A 57-year-old female presented with a lump in the left breast for a month, rapidly progressive. There was no history of breast surgery or breast irradiation. On palpable examination the ill-defined lump of 1.5 cm × 1.0 cms in periareolar region of the left breast was firm, non tender and not attached to the skin or deep structures, and there was no evidence of nipple retraction, skin thickening or axillary lymphadenopathy. Routine blood investigations and a chest x ray were within normal limits. Mammography revealed an ill- defined area of increased soft tissue opacity in the left retro-areolar region without architectural distortion or overlying skin thickening. On USG, there is inflamed fat with heterogenous lesion in the inferior peri-areolar region. Lesion measures 30x28 mm and is predominantly hyperechoic with few central hypoechoic areas. No microcalcification or star shaped opacity is evident. Retro-areolar region is normal. No enlarged lymph nodes are seen in the breast parenchyma. Vascular markings are normal. And was reported as BIRADS III? Fat Necrosis. FNAC was suggestive of low grade spindle cell tumour and the diagnosis was confirmed on core needle biopsy. A wide local excision of lump was performed. The cut surface of the tumour appeared ill defined, spongy and haemorrhagic. Microscopic examination of the mass demonstrated an infiltrative tumour comprising of vasoformative channels, solid areas and spindle cell fascicles. Compressed vascular lumina is lined by atypical oval plump endothelial cells with focal hobnailing. Solid areas show spindled cells with moderate nuclear enlargement and pleomorphism with increased mitotic activity (MF- 6 to 8/HPF). Features were suggestive of grade I angiosarcoma. On immunohistochemistry the tumour was positive for CD31, CD 34 and Vimentin. At two-month follow-up the patient was free of any evidence of recurrence.

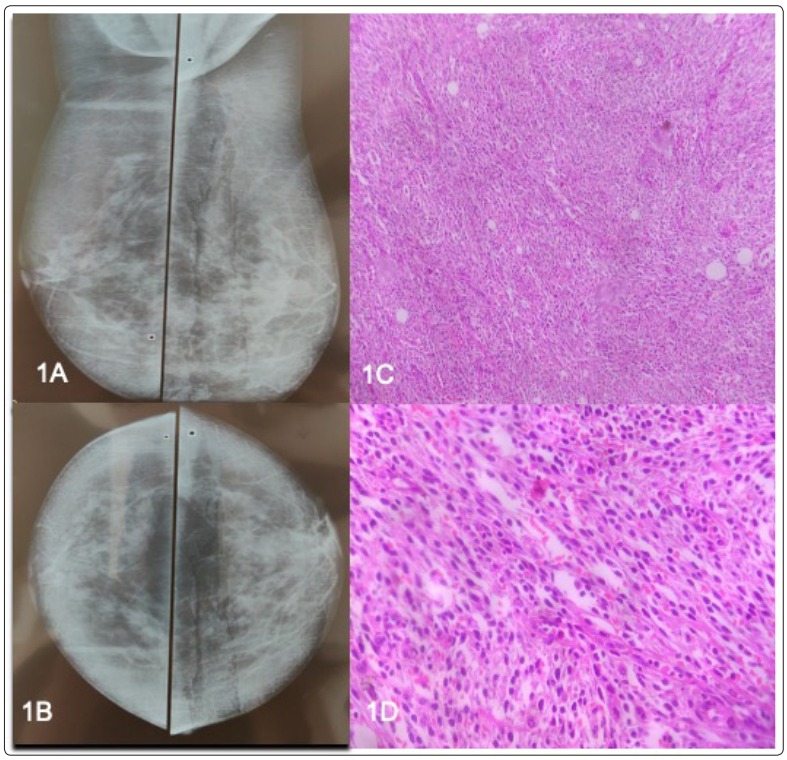

Figure 1 A-B: Mammography showing an ill- defined area of increased soft tissue opacity in the left retro-areolar region without architectural distortion or overlying skin thickening.
Figure 1C: Microphotograph showing infiltrative tumour comprising of vasoformative channels, solid areas and spindle cell fascicles (H and E X100)
Figure 1 D: Solid areas show spindled cells with moderate nuclear enlargement and pleomorphism with increased mitotic activity MF- 6 to 8/HPF (H and E X400)

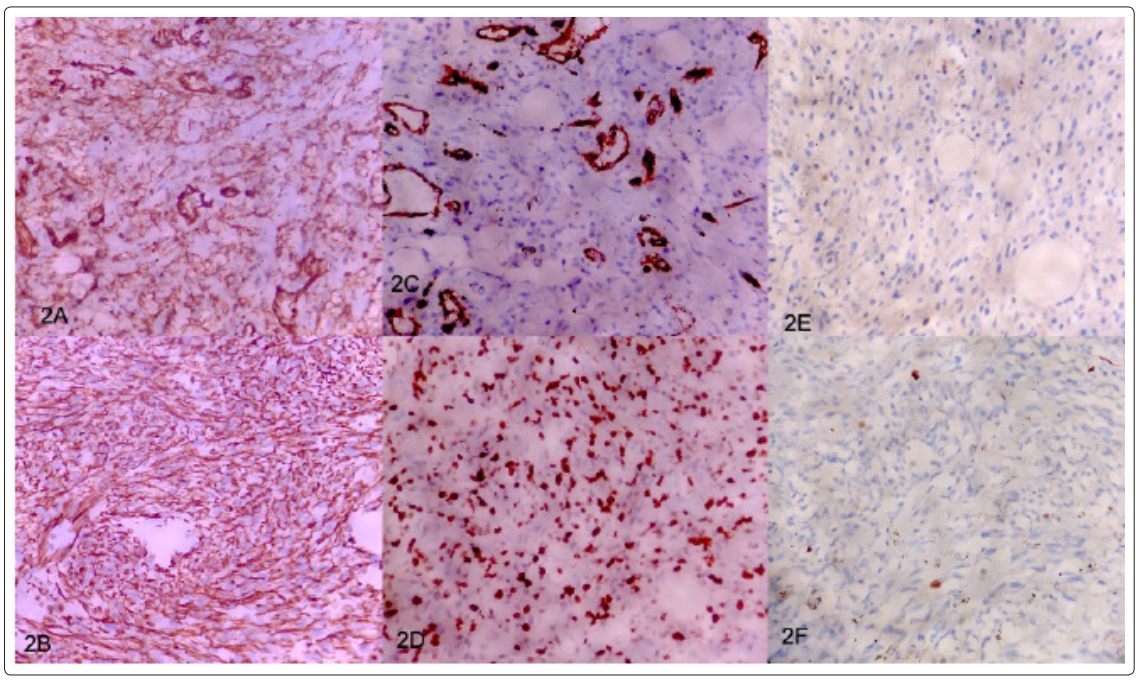

Figure 2 A: Immunoreactive score 4 in tumour cells (CD31 IHC stain X400)
Figure 2B: Immunoreactive score 4 in tumour cells (Vimentin IHC stain X400)
Figure 2C: Immunoreactive score 4 in tumour cells (CD 34 IHC stain X400)
Figure 2D: Percentage of tumour cells with nuclear positivity -70% (Ki-67 , IHC stain X400)
Figure 2E: Non-immunoreactive score 0 in tumour cells (CD99, IHC Stain , X400)
Figure 2F: Non-immunoreactive score 0 in tumour cells (Beta Catenin , IHC stain , X400)

Table 1: Summary of IHC

Antibody	Clone	Interpretation
CD31	GM006	Immunoreactive score 4 in tumour cells
CD34	QBEND10	Immunoreactive score 1+ (patchy) in tumour cells
CD99	12E7	Non-immunoreactive score 0 in tumour cells
Desmin	GM007	Non-immunoreactive score 0 in tumour cells
EMA	E29	Non-immunoreactive score 0 in tumour cells
Ki-67	30-9	Percentage of tumour cells with nuclear positivity -70%
S-100	4C4.9	Immunoreactive score 1+ (patchy) in tumour cells
SMA	1A4	Non-immunoreactive score 0 in tumour cells
Vimentin	V9	Immunoreactive score 4+ in tumour cells
B-Catenin	EP35	Non-immunoreactive score 0 in tumour cells
HHV-8		Non-immunoreactive score 0 in tumour cells

Discussion

Angiosarcoma can occur in any part of the body, most commonly in the skin of head, neck and scalp, however the breast being the rarest site accounting to only 0.05% [3]. Breast angiosarcoma is categorized into primary and secondary due to post- radiation. Secondary angiosarcoma develops in chronic lymphedema, resukting from axillary dissection which is called as Stewart-Treves Syndrome [4]. Mean age of presentation is between 30 and 50 years whilst secondary develops 5 to 10 years after breast cancer treatment.

Primary angiosarcoma of the breast carries poor prognosis. The median disease free survival and OS times are 2.26 and 2.96 years respectively [5]. The prognostic factors includes tumour size, histologic characteristics and surgical margin.

Low grade angiosarcomas are readily mistaken for benign hemangioma. Angiolipoma features intermingling vessels and fat lobules which may be mistaken for invasion of fat by an angiosarcoma. Papillary endothelial hyperplasia as described by Branton et al. is the "great imposter for angiosarcoma" [6]. The treatment of angiosarcoma of the breast is early and complete surgical excision of the mass with tumor-free margins because of the neoplasm often extends microscopically beyond its gross limits. Adjuvant chemotherapy that includes doxorubicin for patients with poorly differentiated angiosarcoma of the breast results in a higher proportion of patients who are relapse-free compared to patients not receiving adjuvant chemotherapy [7]. Radiotherapy is reserved after lumpectomy, and following total mastectomy if the tumor is larger than 5 cm, the margins are positive, or if the skin or regional nodes are affected.

Conclusion

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.

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