Angiosarcoma of Breast in Young Female: A Diagnostic Challenge to Radiologist

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Abstract- Angiosarcoma is a rare cancer that originates from the cells that line the blood vessels or lymph vessels. (1)

Breast Angiosarcoma is of two forms, primary and secondary. The cause is usually unknown in the primary form and the secondary form which is more common is usually associated with a history of irradiated breast. Clinical presentation and the age of presentation of these two forms differ significantly. However, both seem to have a very high malignant potential and a guarded prognosis. (2)

In this case report, we illustrate clinical presentation, imaging features, histopathological features and management of a case of angiosarcoma of the breast in a young female.

Index Terms- Blood vessel lining cells, Imaging features, Primary angiosarcoma, Secondary angiosarcoma

I. INTRODUCTION

Angiosarcoma (AS) accounts for 1% of all soft tissue breast tumors. It can present as primary or as secondary lesions of the breast, secondary lesions being the most common presentation among those having a past history of irradiation to the breast.

Primary AS is commonly seen in women during the fourth to sixth decade of life, usually coming with complaints like palpable mass, increase in breast size or reddish discoloration of the skin. Secondary AS is seen in older age group women during the seventh to eighth decade of life, who usually present approximately after 10-11 years following radiotherapy for breast cancer.

Primary variant arises in the breast parenchyma typically, with or without overlying skin involvement. It accounts for less than 0.04% of all malignant neoplasms in the breast.

The first ever case of Primary and secondary breast AS was presented by Borrman in 1907 and Body et al in 1987 respectively. Prognostic factors and management methods have not yet been clearly described because of the rarity of the condition. (2)

In this literature, we review the clinical presentation, Imaging findings and histopathological features in a case of Primary Angiosarcoma of the breast.

Clinical presentation:

In our case, we had a 17-year-old female who came to the surgery OPD with complaints of pain and increase in size of the right breast for 2-3 months. The size of the breast was increasing gradually with few dilated veins and reddish discoloration of the skin over the right breast. On physical examination, Right breast showed a large palpable mass around 10x15cm which was firm in consistency. There was no evidence of discharge from the nipple, no nipple retraction. Both axillae were normal on examination. No significant past history. Clinically, a diagnosis of mastitis of Tubercular etiology was made, however, the mass did not subside and cytology did not show epithelioid cells and hence further evaluation was suggested.

Evaluation and diagnosis:

Bilateral sonomammogram was done as the first line of investigation for the patient, after which MRI was suggested. Patient's platelet count was seen to be dropping, however, rest of the blood and other Laboratory parameters were within normal limits. MRI was repeated after 1 month of initial presentation for reassessment and pre-operative planning and there was a significant increase in the size compared to the previous study. Ultrasound-guided FNAC of the lesion was done and the results suggested a highly vascular tumor. Following which wide bore biopsy was done. Based on radiological and pathological findings, a diagnosis of angiosarcoma was given. The patient underwent a total right mastectomy. The Immunohistochemistry (IHC) reports showed cells positive for CD 31 and CD 34. the diagnosis of Angiosarcoma was confirmed.

II. IMAGING FINDINGS

Bilateral Sonomammogram:

Sonomammogram done in Philips Affiniti 70 machine demonstrated a large heterogenous lesion involving all the quadrants of the right breast. The lesion showed increased vascularity on doppler with multiple whorls and multiple venous spaces with slow flow within. Normal breast parenchyma could not be delineated. Left breast and bilateral axilla were normal.

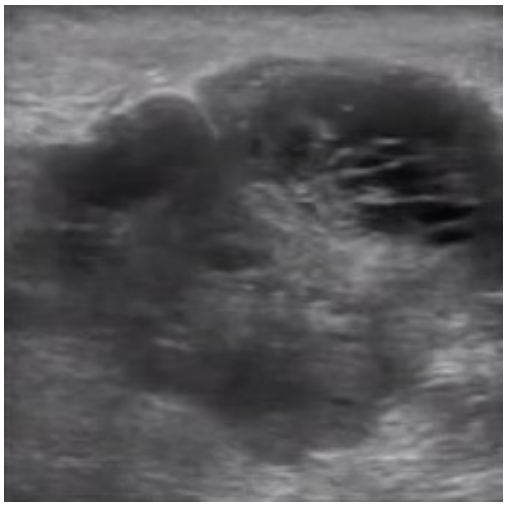
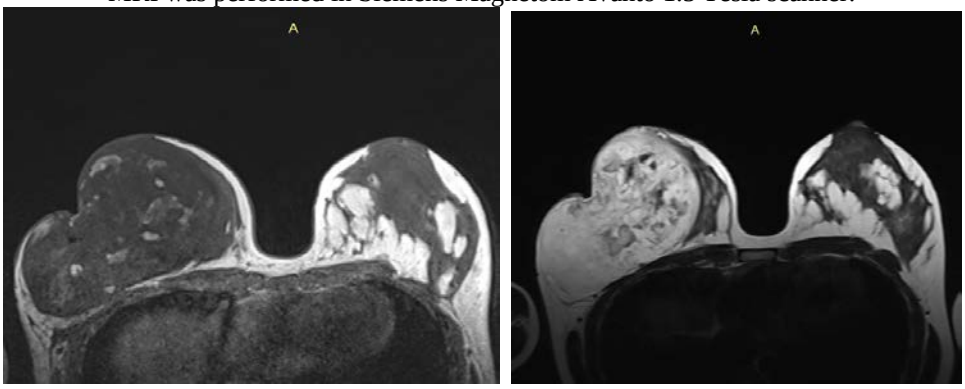


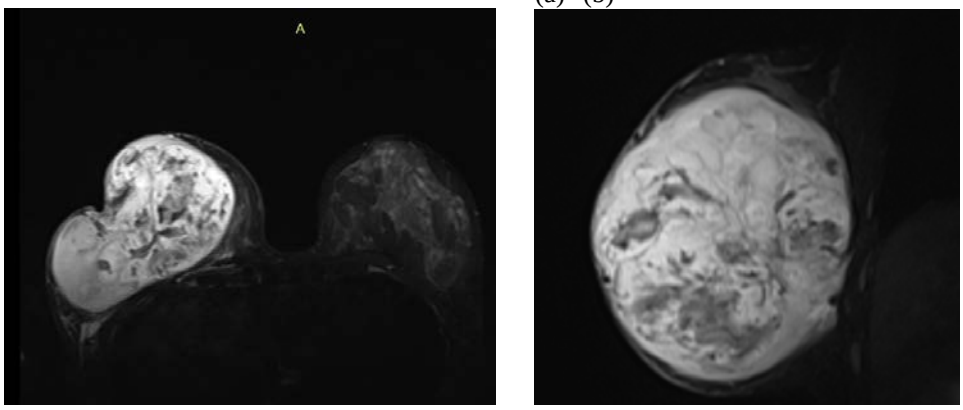
Image.1: Grey scale image demonstrating a fairly well-defined heterogenous lesion with multiple vascular spaces within

MRI FINDINGS:

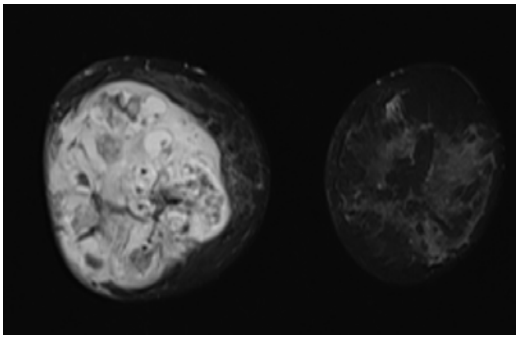
MRI was performed in Siemens Magnetom Avanto 1.5 Tesla scanner.



(a) (b)



(c) (d)



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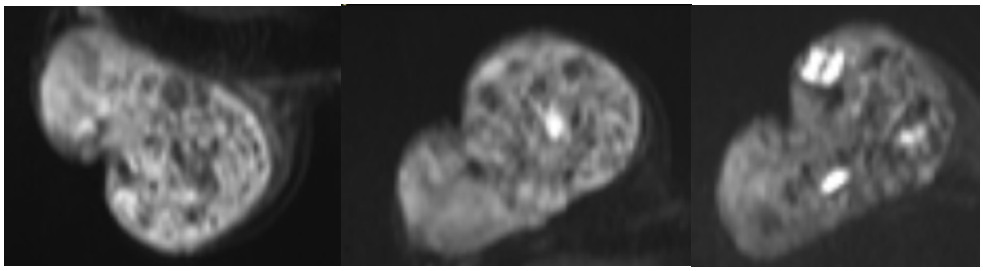

(f)

Image.2: MRI of right breast showed a large well encapsulated heterogeneously enhancing lesion.(a)Axial T1W image showing heterogeneously hypointense lesion.(b)&(c)Axial T2W and STIR images showing heterogeneously hyperintense lesion with multiple haemorrhagic and cystic areas within.(d)&(e)Sagittal and coronal T2 W images showing fluid-fluid levels ,slit like vascular spaces and flow voids within.(f)Axial diffusion weighted images showing multiple ill-defined areas of diffusion restriction.

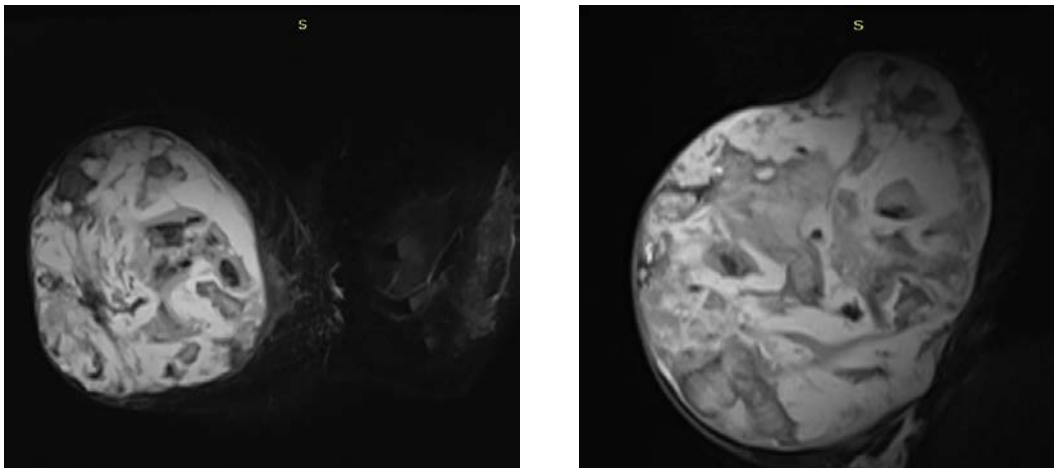

Image 3: Axial and Coronal T2 W MR images of the same patient in a study done after 1 month. There is significant increase in the size of the lesion compared to the previous study. Large well encapsulated solid-cystic mass with internal vascular spaces is seen.

III. HISTOPATHOLOGICAL FINDINGS

Fine needle aspiration cytology was advised for the patient in order to rule out a possibility of malignancy. The specimen received by laboratory consisted of single pale brown tissue measuring 2.5x1. 5x1cm.Histopathological examination showed micropapillary fronds lined by epithelial and myoepithelial cells supported by fibrovascular stroma. Adjoining tissue showed Pseudo-angiomatous stromal hyperplasia like stroma and organizing hematoma. Immunohistochemical studies revealed a positive reaction of the neoplastic cells for; CD34, CD31, and the results appeared negative for Pan CK,Her 2 neu . The patient underwent Radical modified mastectomy for the same and the received mastectomy specimen measured 14.5x10x4.5cm, solid and cystic areas are seen with multiple papillary projections. The lesion was away from the nipple-areolar complex and resected margins were free from the lesion. On microscopy, the tumor composed of cystic areas filled with necrotic and hemorrhagic material and lined by endothelial cells arranged in papillae with complex branching pattern. Endothelial cells are spindle to epitheloid having a hyperchromatic nucleus

with significant pleomorphism. Tumor cells were arranged in solid nests and focal cribriform spaces having a pleomorphic hyperchromatic nucleus. Solid areas showed focal atypical mitosis(10-12/10hpf). Many areas showed capillary and cavernous hemangioma-like areas. More than half of the tumor cells composed of necrosis. Focal areas of blood vessels, papillary projections, and the hyperplastic epithelium is noted. Nipple-areola complex is free from the tumor.

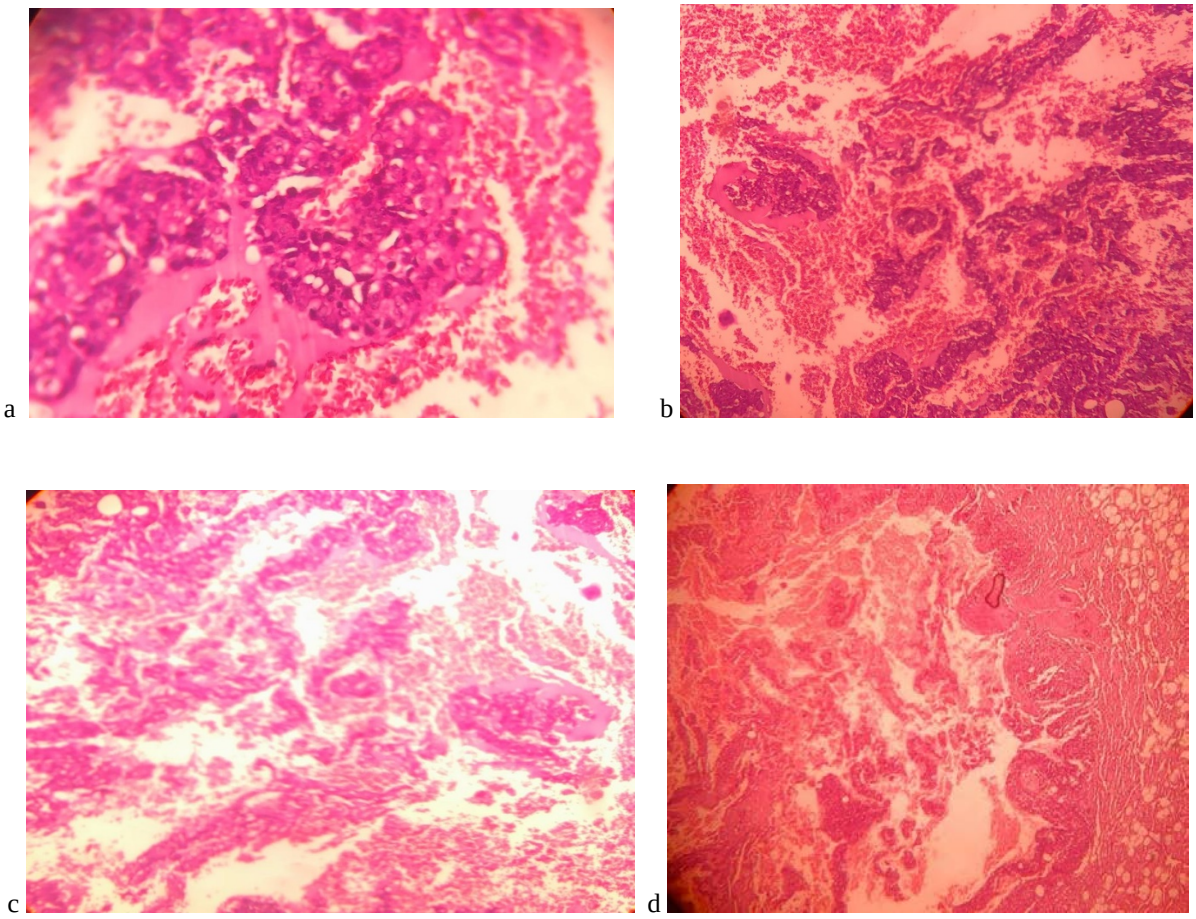

Image 4: Slides were air-dried and fixed in alcohol and stained by Giemsa and Papanicolaou stain.a)&b)Tumor composed of cystic areas filled with necrotic and hemorrhagic material and lined by endothelial cells arranged in papillae with complex branching pattern. c)&d)Endothelial cells show hyperchromatic nucleus with significant pleomorphism.Tumor cells are seen arranged in solid nests and focal cribriform spaces having pleomorphic hyperchromatic nucleus.

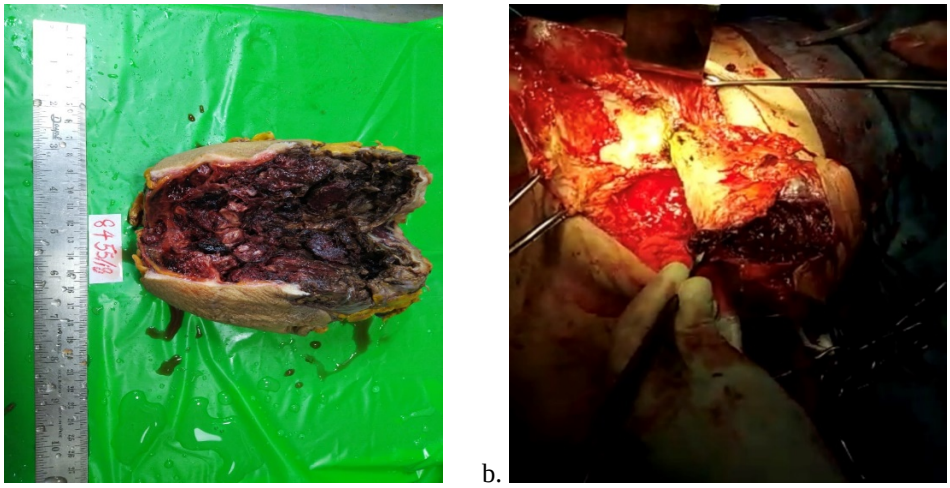

Image 4: a) Gross specimen of right breast shows Grey brown areas with focal grey white areas. Solid and cystic components are seen with multiple papillary projections. Areas of haemorrhage and necrosis are also noted. b)Intra-op image during right modified radical mastectomy shows a highly vascular tumor.

IV. Discussion

Angiosarcomas are tumors arising from an endovascular origin. These are rare tumors which can arise from any organ in the body.

Although rare, breast is one of the favorable sites for angiosarcoma with an incidence of approximately 8%.

Until now 187 cases of angiosarcoma have been reported. Besides the head, thigh, and arm, the breast is one of the more common sites for angiosarcoma. However, only 0.04% of malignant breast tumors have been reported to be angiosarcoma. (3)

Primary breast angiosarcomas occur most commonly in women between the ages of 20-40 years, and typically present as an ill-defined breast mass. Secondary angiosarcomas usually occur in older women, following the treatment of breast cancer. They can arise in the lymphedematous upper extremity or in the chest wall, following radical mastectomy and irradiation, or in the breast parenchyma itself following a breast-conservative treatment.

Because of its rarity and its deceptive benign histologic appearance, angiosarcomas of the breast are often misinterpreted as benign lesions at initial biopsies.

There is no significant characteristic mammographic pattern for angiosarcoma, however, the literature shows varied appearances like ill-defined breast masses without spiculations and calcifications which are common in other breast malignancies. Even on sonomammogram, Angiosarcomas have variable features such as well-circumscribed or poorly marginated , hypoechoic or mixed echoic masses. Depending on the presence of vascular channels and cellular components, the heterogeneity of the lesion differs.

Unlike most of the other malignant breast lesions, angiosarcoma does not show angular margins and posterior shadowing. On color Doppler ultrasound, they show increased vascularity within the lesions with multiple vascular channels. The ultrasound and mammography appearance of angiosarcoma are non-specific. Magnetic imaging resonance (MRI) examination is a better diagnostic tool in breast angiosarcoma. Low-grade angiosarcoma presents as a large lobulated mass with indistinct borders that shows hypointense signal on T1-weighted images and hyperintense on T2-weighted images, high-grade angiosarcoma demonstrates hyperintensity on both T1 weighted and T2 weighted images, with mixed intensity foci within the tumor which is contributed by hemorrhage or venous lakes. Aggressive breast angiosarcomas show rapid post contrast enhancement and washout pattern. (4)(5)(6)(7)(8).

Histopathologically, tumors characteristically show anastomosing vascular channels with neoplastic and proliferative endothelial cell lining, with mild cytologic atypia and rare mitotic figures. Solid areas are composed of numerous large neoplastic cells arranged in sheets without characteristic blood channels, and frequent mitotic figures with high-grade tumors showing large areas of necrosis. Immunohistochemical studies play an important role in differentiating angiosarcoma from other poorly differentiated carcinomas, soft tissue sarcomas, and malignant melanomas. In well-differentiated angiosarcomas, neoplastic cells have a strong positive reaction for CD34, CD31, vimentin, and factor VIII-related antigens.

Angiosarcoma is a very aggressive tumor with a poor lookout, a high rate of local recurrence, and multiple-organ involvement, especially in the lungs, liver, regional lymph nodes, and bones (10). Prognosis depends on the tumor histologic grade.

Treatment of angiosarcoma is challenging owing to the unpredictable behavior of the tumor. However, wide local excision, Modified radical mastectomy or mastectomy followed by adjuvant chemotherapy have shown acceptable outcomes(4),(9). Dissemination of angiosarcoma happens via the hematological route, therefore chemotherapy after surgery, especially for high-grade tumors, or in cases of distant metastasis, is seen to be beneficial (2).

Perilobular hemangioma which is a benign lesion is the differential diagnosis that should be borne in mind for angiomatous lesions of the breast.

V. Conclusion

As we know from literature, the incidence of primary angiosarcoma is rare. However, it should be considered as a differential diagnosis with fibroadenoma or phyllodes tumor. High-grade angiosarcomas may show similar histopathological features as other soft tissue sarcomas, in such cases, immunohistochemical (IHC) studies are useful for confirming the diagnosis. (11)

Although a diagnostic pitfall has been repeatedly emphasized in some of the previous reports, the problem of underdiagnosing still persists in recent years. In a few of the cases, there was a delay of up to two years in recognizing the malignant nature of the tumor.

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