



PRIMARY ANGIOSARCOMA OF THE BREAST-MISTAKEN YET RARE AGGRESSIVE NEOPLASM

Radio-diagnosis

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ABSTRACT

Primary angiosarcoma of the breast is a rare potentially life-threatening disease. Given its variable and non-specific clinical, radiological and pathological presentation, accurate diagnosis is a challenge. Primary angiosarcoma of the breast predominantly occurs in younger patients and it is often overlooked and misdiagnosed in radiology and pathology. To ensure that this aggressive malignancy is not overlooked, we need to be aware of the fact that such tumours may present with non-specific imaging features. Initially, it was diagnosed as Pseudo-angiomatous stromal hyperplasia (PASH) with radio-pathological discordance, then IHC and second HPE led to diagnosis of angiosarcoma. Hence we need to be aware of its imaging presentation. We hereby are presenting Mammographic, Ultrasound, doppler, PET CT and MRI findings of a patient who presented to our hospital with a large lump in the right breast.

KEYWORDS

Angiosarcoma; Breast; Ultrasound; MRI; PET CT; Doppler

CASE REPORT

A 57 yrs old female patient Parity index-P2L2 with known case of hypertension and diabetes since 1 year presented with a history of lump in the right breast which she noticed incidentally 4 months back. No history of hormonal intake or family history of breast malignancies. On clinical examination swelling of 5cm horizontally and 4cms vertically was palpated at 3 'o clock – 5 'o clock position (lower inner quadrant) of right breast. Oval in shape irregular margins, hard in consistency not fixed to skin or chest wall. No axillary lymph nodes.

Imaging Findings:

Mammogram shows predominantly fibrofatty pattern composition of Breast. Right breast showed a fairly lobulated radio dense mass lesion in the lower inner quadrant extending to retro areolar region, the mass showed Focal microlobulations and spiculations. No lymphadenopathy /microcalcifications/ skin thickening noted figure (1a&1b). Left breast was normal.

Ultrasound of the right breast showed lobulated mixed echogenic mass lesion of size 3.5 X 2.2cms. Wider than taller seen at 2-4 o clock position extending to retro areolar. Focal Multiple micro lobulations and cystic areas noted within the lesion. Perilesional halo disrupted, Low resistance vascularity doppler. Satellite nodule seen adjacent to the lesion. No evidence of axillary lymphadenopathy (figure 1c-1f).

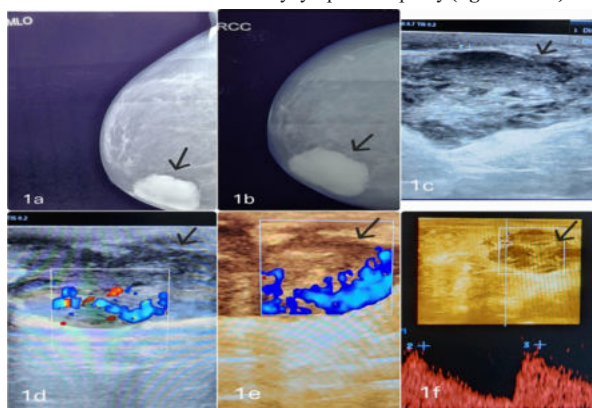


Figure 1- Mammography and USG of right breast showing mass lesion; 1a (MLO view) & 1b (CC view) predominantly fibrofatty breast shows a fairly lobulated radio dense mass lesion (black arrow) in the lower inner quadrant extending to retro areolar region, the mass showed Focal micro lobulations and spiculations. No lymphadenopathy /microcalcifications/ skin thickening noted. 1c-1f USG images shows a lobulated mixed echogenic mass lesion of size 3.5 X 2.2 cms (black arrow). Wider than taller seen at 2-4 o clock position extending to retro areolar. Focal Multiple micro lobulations and cystic areas noted within the lesion. Perilesional halo disrupted, Low resistance vascularity doppler.

MRI breast showed a 5.2 x 4 x 3.5 cms heterogenous lesion with spiculated border and disrupted perilesional halo in the inner aspect of right breast extending to the retro areolar region. One component extends to subcutaneous plane. The lesion appeared hypointense on T1W with focal hyperintensity seen at periphery possibly haemorrhage and Heterogenous predominantly hyperintense with multiple areas of hypo intensity within it on T2 w and STIR sequences (figure 2a-2c).

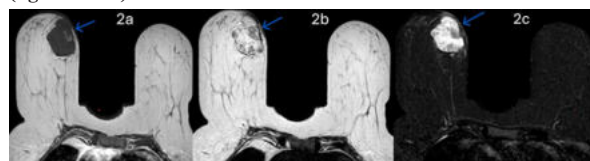


Figure 2: On MRI breast shows a T1w T2w & STIR shows a 5.2 x 4 x 3.5 cms heterogenous lesion (blue arrow) with spiculated border and disrupted perilesional halo in the inner aspect of right breast extending to the retro areolar region. One component extends to subcutaneous plane, the lesion appeared hypointense on T1W (2a) with focal hyperintensity seen at periphery possibly haemorrhage and Heterogenous predominantly hyperintense with multiple areas of hypo intensity within it on T2W (2b) and STIR(2c) sequences.

Dynamic contrast imaging- Rapid uptake with Persistent and progressive enhancement (figure 3a-3b, 3d & 3h).

DWI /ADC- Lesion appears bright on DWI (figure 3c) with restricted diffusion only in the peripheral aspect with high ADC value (1.2×10^{-3}) (figure 3e-3f).

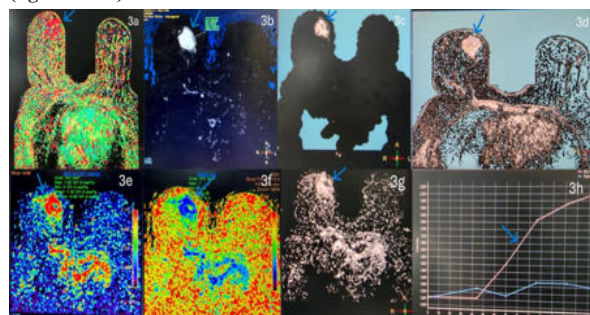


Figure 3: T1W perfusion imaging (3a&3b), Dynamic contrast enhancement imaging (3d) and dynamic contrast/kinetic curve (3h) showed a lesion in the right breast with Rapid uptake with Persistent and progressive enhancement. In DWI (3c) mass lesion appears hyperintense with Slight peripheral restriction on ADC images (3e-3g) with ADC value of 1.2×10^{-3}

Final diagnosis includes possibilities of high-grade neoplasm with intense vascularity without ductal involvement likely sarcoma needs

HPE and IHC correlation -BIRADS 5 lesion.

On HPE the Initial FNAC report came as Pseudo-angiomatous stromal hyperplasia (PASH), but biopsy reported as Pseudo-angiomatous stromal hyperplasia (PASH)/Angiosarcoma- and suggested IHC which came IHC- Triple negative, CD31 & CD34 positive which is endothelial markers. Final diagnosis was made as Angiosarcoma (figure 4a,4b&4c)

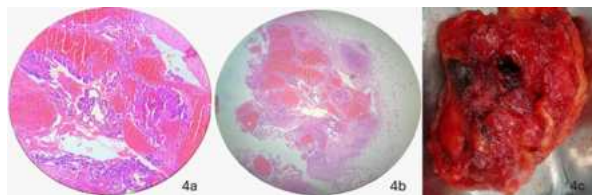


Figure 4: Microscopy (4a&4b) and gross specimen (4c) showed vascularised lesion with many capillary sized vessels arising from medium sized vessels which are lined by plump endothelial cells which are hyperchromatic at places and shows hob nailing, focally well-formed capillary is also seen. Large areas of haemorrhage and focal areas of coagulative necrosis are seen. Many of these capillaries show hyalinisation. Adjacent tissue shows fibro collagenous tissue along with adipose tissue. These capillaries are seen infiltrating the adjacent stroma focally. One of the section shows benign duct lined by bilateral epithelium.

The PET CT was suggested to look for distant metastasis showed uptake of FDG in the lobulated mass with mass showing persistent enhancement in delayed scans. No evidence of any axillary nodes or distant metastasis (**figure 5a-5c**).

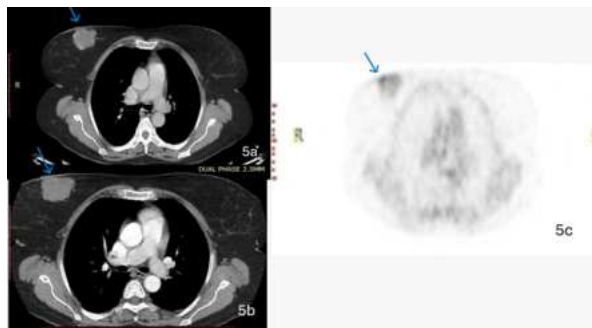


Figure 5: PET CT (5a,5b&5c) showed uptake of FDG in the lobulated mass with mass showing persistent enhancement in delayed scans. No evidence of any axillary nodes or distant metastasis.

DISCUSSION AND CONCLUSION:

Angiosarcomas are aggressive tumors arise from vascular structures and often have metastasis at the time of diagnosis.¹ They are very rare and account for < 1% of all sarcomas, and in breast extremely rare <0.04% of all primary malignant breast tumors.² It can be categorised into primary and secondary breast angiosarcomas. Primary angiosarcomas of the breast occur sporadically in young women during 3-4 decade and usually present as palpable masses. Secondary angiosarcomas occur most frequently after breast conservation therapy with radiation therapy; the average latency period is 5-6 years.³ The prognosis is poor hence early adequate diagnosis is warranted. There is no pathognomonic signs on single radiological investigation, Multidisciplinary approach characterizing these tumors and raising the suspicion of a vascular tumor.

On mammography, Primary angiosarcomas of the breast appears as a not circumscribed mass and lacking the spiculations usually seen in ductal origin breast cancer, rarely manifest coarse, nonbranching microcalcifications.⁴ Many cases have shown that they are not picked in mammograph predominantly in young female due to dense breast tissue.^{1,2}

On Sonography the lesion is seen a solid mass that may have well-defined or lobulated margins with no posterior acoustic shadowing and sometimes as a complex cystic and solid areas.⁵ Studies shows that Primary angiosarcomas of the breast were all hyper vascular on colour Doppler with low resistance vascularity.¹

MRI is the best imaging modality for its characterization, MRI

findings showed T1WI low signal with focal areas of high-signal-intensity due to areas of haemorrhagic nature or intra-tumoral hematoma, T2WI usually has high signal intensity. Slow flowing blood in a large anastomotic vascular space seems to cause high signal intensity in T2WI. When this structure forms a venous lake, it shows a cystic cavity. In our study DCE showed a rapid enhancement pattern with persistent and prolonged enhancement in delayed scans similar to few studies reported like Wang et al.Breast J 2017.⁶ Few other studies showed type 1 and Type 3 kinetics.⁷ Other features like hyperintense on Diffusion weighted study with slight restriction on ADC.⁸

Our case has all of the characteristic MRI findings of Primary angiosarcomas of the breast. PET-CT is useful for confirming preoperative axillary involvement and distant metastasis.

The prognosis of the Primary angiosarcomas of the breast is similar to other histopathology type of sarcoma and is related to the tumor size, grade, and the status of resection margin,⁹ a Surgical removal followed by adjuvant chemotherapy seems to improve the patient prognosis.¹⁰ However, the overall prognosis is poor compared to that of mammary carcinoma. Total mastectomy is mandatory because it often invades microscopically beyond its gross limits, and axillary lymph node dissection is not necessary since nodal metastasis is rare.

In our case report it was a low grade Primary angiosarcomas of the breast without any local or distant metastasis hence total mastectomy was performed without any adjuvant chemotherapy or axillary lymph node dissection.

Although rare for breast masses with increased vascularity, Angiosarcoma must be kept as differential, T2 hyper intensity, Any type of DCE curve predominantly type I, bright on DWI/ADC, High ADC values, Cystic areas with low resistance vascularity on USG/Doppler, absence of lymphadenopathy inspite of large mass should raise the possibility of Primary breast angiosarcoma.

Abbreviation

- PASH: Pseudo-angiomatous stromal hyperplasia
- USG: Ultrasound
- MRI: Magnetic resonance imaging
- PET CT: Positron emission tomography and computed tomogram
- IHC- Immuno-histochemistry
- HPE: Histopathology examination
- STIR: short tau inversion recovery
- DWI: Diffusion weighted imaging
- ADC-Apparent diffusion coefficient
- DCE-Dynamic contrast enhancement

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