

## Primary Angiosarcoma of the Breast in an Adolescent Female

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### 1. Abstract

#### 1.1. History

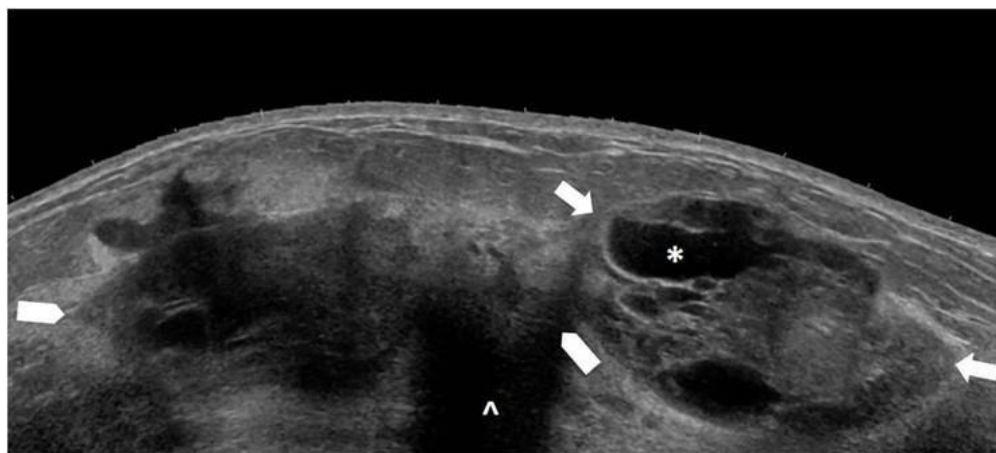
A 17-year-old female presented to the emergency room with palpable right breast masses. On physical examination, there was diffuse ecchymosis of the right breast. A firm mobile mass was present in the upper outer and upper inner quadrant of the right breast. Due to the patient's age, a diagnostic US (Figure 1) was initially performed [1]. Afterwards, she underwent a diagnostic mammogram (Figure 2).

#### 1.2. Imaging Findings

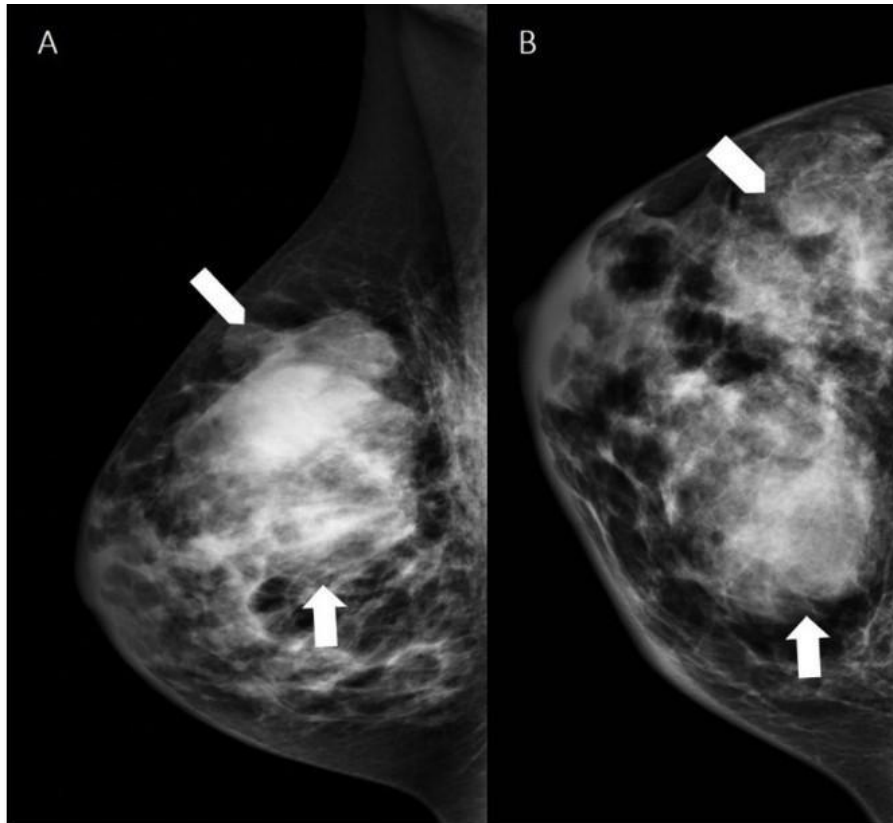
Ultrasound of the right breast shows a 6.5 cm, irregular, heterogeneous mass with non-circumscribed margins and mixed posterior features within the upper outer quadrant. A 5.5 cm, oval, mixed solid and cystic mass, with circumscribed margins and parallel orientation is present within the upper inner quadrant (Figure 1). The mammogram demonstrates a 6.5 cm, irregular, isodense mass with indistinct margins within the upper outer

quadrant and a 5.5 cm, oval, isodense mass with indistinct margins in the upper inner quadrant (Figure 2). The left mammogram is normal (not shown). An US-guided biopsy of the two suspicious right breast masses were subsequently performed. Histopathology shows a primary angiosarcoma with high grade nuclear atypia and numerous mitotic figures at both sites (Figure 3). The patient then underwent a bilateral breast MRI with contrast.

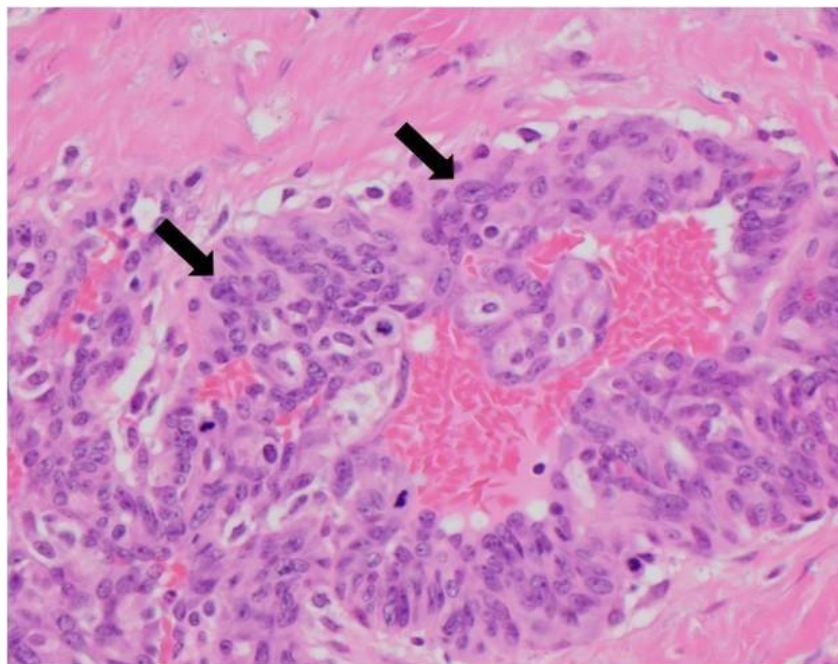
The MRI demonstrates an asymmetrically enlarged right breast with two large masses that have heterogeneous T2 signal. The post-contrast images demonstrate a 6.5 cm, irregular mass with rim enhancement within the upper outer quadrant and a 5.5 cm, oval mass with rim enhancement in the upper inner quadrant of the right breast (Figure 4). The left breast is normal on MRI. The patient then underwent staging with F-18 FDG PET-CT, which shows increased heterogeneous rim uptake of the FDG involving the right breast masses (Figure 5). There was no distant disease.



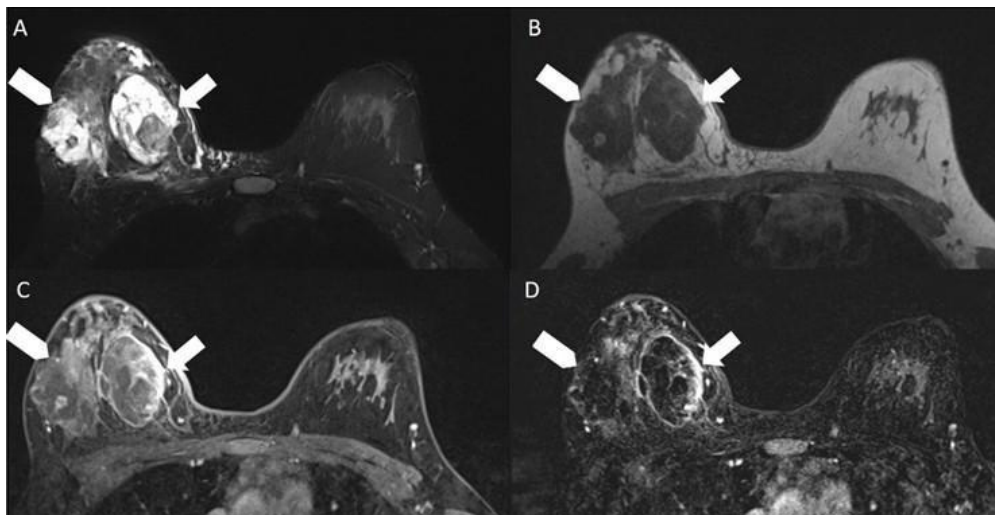
**Figure 1:** Large field-of-view ultrasound of a 17-year-old female with palpable right breast masses. The image shows a 6.5 cm, irregular, heterogeneous mass (arrowheads) with non-circumscribed margins and mixed posterior features with shadowing (^) within the upper outer quadrant. In addition, a 5.5 cm, oval, mixed solid and cystic (\*) mass (arrows), with circumscribed margins and parallel orientation is present within the upper inner quadrant.



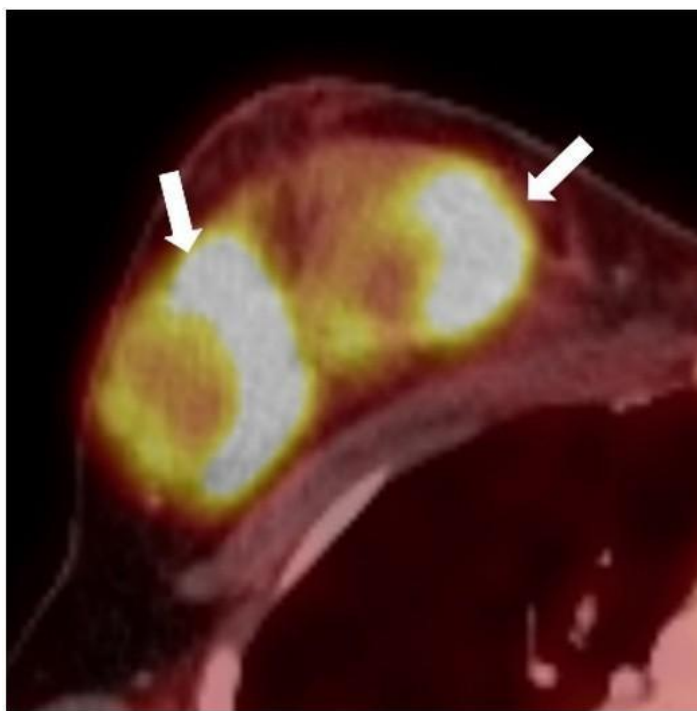
**Figure 2:** Mammographic images of a 17-year-old female with palpable right breast masses. Mediolateral oblique (A) and craniocaudal (B) mammogram images demonstrate a 6.5 cm, irregular, isodense mass with indistinct margins within the upper outer quadrant of the breast (arrowhead). A 5.5 cm, oval, isodense mass with indistinct margins is also present in the upper inner quadrant of the breast (arrow). The left mammogram is normal (not shown).



**Figure 3:** Histopathology of the right breast. Primary angiosarcoma with high-grade nuclear atypia (arrows) is present. Hematoxylin and eosin stain, 40x.



**Figure 4:** Bilateral breast MRI of a 17-year-old female with palpable right breast masses. Pre-contrast non-fat suppression axial T2 (A) and axial T1 fat suppression (B) demonstrate an asymmetrically enlarged right breast with two large masses (arrows) that have heterogeneous T2. The post-contrast axial T1 image (C) and post-contrast axial T1 subtraction image (D) demonstrate a 6.5 cm, irregular mass with rim enhancement (arrowhead) within the upper outer quadrant and a 5.5 cm, oval mass with rim enhancement (arrow) in the upper inner quadrant of the right breast. No nodal involvement is seen. The left breast is normal.



**Figure 5:** Axial fused F-18 FDG PET-CT demonstrates increased heterogeneous rim uptake of FDG involving the right breast masses (arrows). There was no evidence of metastatic disease.

### 1.3. Differential Diagnosis

The differential diagnosis for a mixed solid and cystic mass in a young female patient include infection and hematoma/trauma in the appropriate clinical setting. Less common etiologies to consider include an infarcted fibroadenoma, papilloma, phyllodes tumour, invasive ductal cancer and primary breast angiosarcoma.

### 1.4. Discussion

Primary angiosarcoma (PAS) is a malignancy of breast endothelial origin that is not related to prior radiation and has an incidence of 0.04% [2,3]. It typically presents in younger women as a palpable mass and 20% may have overlying skin changes,

bluish discoloration [2,4-6].

The imaging findings of PAS are nonspecific. On mammography, it may present as a noncalcified mass with circumscribed or indistinct margins [2,4-6]. Thirty-three percent of PASs have been reported to be occult except for minor skin thickening [2,6]. The sonographic ranges from a hypoechoic to heterogeneous mass with circumscribed or not circumscribed margins to a region of mixed echogenicity [2,4-6]. PAS may be hyper vascular on Power Doppler [2,6]. On MR, it may have high-signal intensity on T2-weighted images with variable kinetics depending on tumour grade [2,4-6]. Due to its nonspecific appearance, needle core biopsy is needed for a definitive diagnosis.

The treatment of PAS is surgical resection with negative margins [2,6]. Nodal involvement is rare since metastases are hematogenous [2,6]. Neoadjuvant or adjuvant chemoradiation therapy have been proposed [2, 6]. The reported overall 5-year survival rate for PAS is 49% [7].

The patient underwent neoadjuvant chemoradiation therapy followed by a right mastectomy. Final histopathology showed a 12.2 cm PAS.

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