Surgery Research Journal
ISSN: 2768-0428

Case Report

Primary Angiosarcoma of The Breast : A Case Report of a Rare Presentation

Obaid Al-Harbi[1], Ahmad Essam Al-Mulla[2], Waleed Bader Abushanab[3], Asmaa Abdulkarim AlSamhan[4]

[1]Consultant Surgeon, Department of Surgery Farwaniya Hospital, Ministry of Health Kuwait (MOH), Kuwait
[2]Consultant General and Bariatric Surgeon, Department of Surgery Farwaniya Hospital, Ministry of Health Kuwait (MOH), Kuwait
[3]Surgical Registrar Department of Surgery, Department of Surgery Farwaniya Hospital, Ministry of Health Kuwait (MOH), Kuwait
[4]Senior House Officer General surgery (SHO), Department of Surgery Farwaniya Hospital, Ministry of Health Kuwait (MOH), Kuwait

Correspondence
Ahmad Essam Al-Mulla MD, FACS
Sabah Al-Nasser, Block 6, P. O. Box 13373, Farwaniya 81004, Farwaniya, Kuwait
Tel. 0096524888000.
Email: draalmulla2007@gmail.com

- Received Date: 23 Aug 2023
- Accepted Date: 29 Aug 2023
- Publication Date: 01 Sep 2023

Abstract
Primary angiosarcoma of the breast is a rare entity, accounting for less than 0.1% of breast malignancies, with potential life-threatening outcomes. Diagnosis of angiosarcoma can be difficult due to its variable and unpredictable clinical, radiological, and pathological presentation. Primary angiosarcoma occurs predominantly in younger populations with no previous risk factors; in contrast, secondary angiosarcoma occurs predominantly in the elderly with a previous of radiation exposure. We describe the case of a 50-year-old female presenting with a large, advanced breast tumour diagnosed as primary angiosarcoma..

Background

Breast angiosarcoma (AB) is a sporadic and progressive endovascular malignant tumour [1], accounting for less than 0.1% of all breast malignancies [2]. This angiosarcoma may be classified as primary or secondary. Primary angiosarcoma arises in non-irradiated breasts and has no previous risk factors; secondary angiosarcoma arises from the dermal and subcutaneous layers of the skin secondary to radiation [3]. Angiosarcoma usually presents as rapidly growing, palpable, painless masses. Here we describe the course and management of a rare progressive primary angiosarcoma in a 50-year-old female with no previous history or risk of radiation exposure.

Case presentation

A 50-year-old female presented to our surgical emergency department, complaining of a one-year history of a right breast mass. The patient had no medical and surgical history and no family history of breast disease or previous radiation exposure. She was married with two children. The mass was progressively increasing in size and was associated with itchiness and pain. The patient had noticed changes in the overlying skin colour and a discrepancy in the sizes of the breasts (Figure1). Vital signs were normal upon examination; breast examination revealed a large fungating mass at the right upper quadrant; the mass was fixed to the chest with bluish discolouration of the breast and enlarged axillary lymph nodes bilaterally.

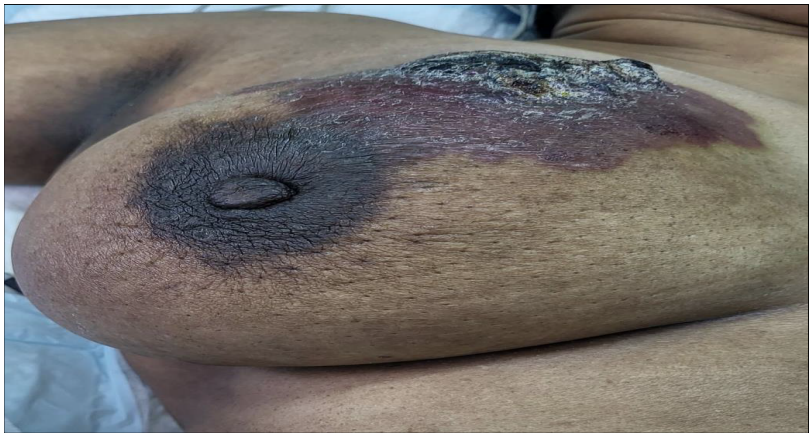

Figure 1. *Right breast angiosarcoma with fungating bluish discoloration mass.*

Citation: Al-Harbi O, Al-Mulla AE, Abushanab WB, AlSamhan AA. Primary Angiosarcoma of The Breast: A Case Report of a Rare Presentation. Surg Res J. 2023;3(2):1-3

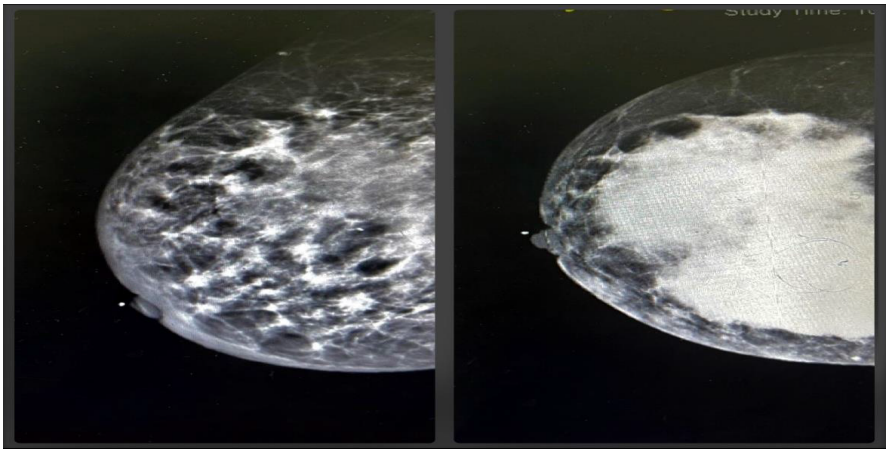

Figure 2. *Mammogram revealing hypoechoic 5.2 x 3.4 angiosarcoma of the breast.*

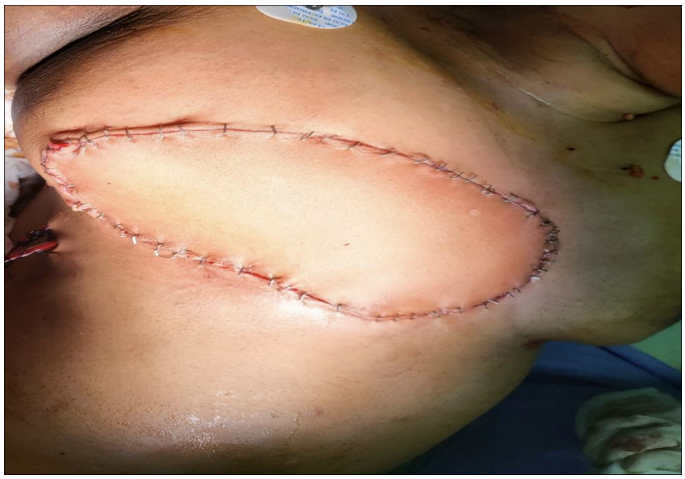

Figure 3. *Radical mastectomy with latissimus dorsai flap reconstruction.*

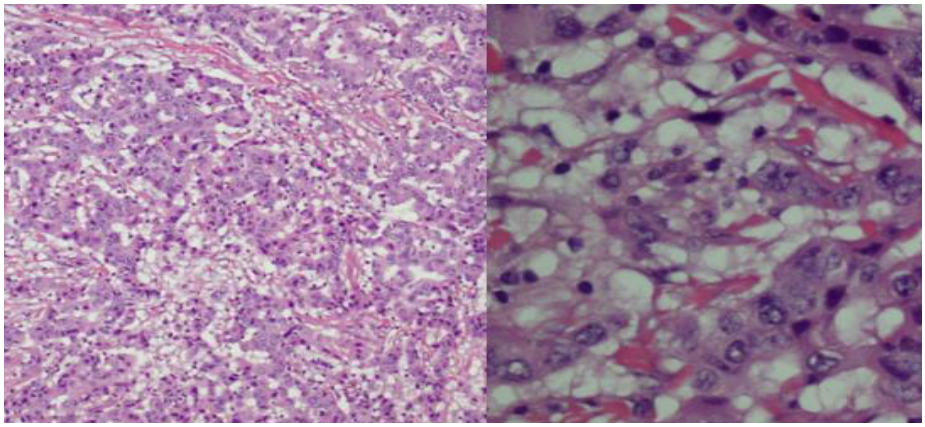

Figure 4. *Histopathology revealing magnified angiosarcoma and normal breast tissue.*

The patient was referred for a mammogram and core biopsy of the mass for further evaluation. Mammography showed a fungating hypoechoic mass occupying the entire breast, mostly retro-areolar. It was 5.3 x 3.4 cm in diameter, with increased vascularity and diffused oedema (BI-RAD 5) (Figure 2). Computer tomography scan of the chest and abdomen showed no signs of metastasis. Histopathology from the core biopsy showed atypical cells with thick collagen bundles and entrapped adnexal structures, strongly positive for CD-31 and CD-34 on the immunochemical stain, indicative of atypical vascular proliferation.

After an anaesthesiology evaluation for surgery , the patient was informed of the finding and consented to a radical mastectomy and latissimus dorsai flap reconstruction by the plastic surgery team immediately (Figure 3). Histopathology of the specimen confirmed the angiosarcoma diagnosis.

The patient had an uneventful post-operative recovery and was discharged six days after surgery and completing antibiotics. She was referred to medical oncology in her origin country for further evaluation and was lost to follow-up three months after surgery.

Discussion

Angiosarcoma of the breast is an infrequent tumour. Of the two types, primary and secondary, primary angiosarcoma of the breast (PAB) is the rarest, accounting for only 20% of angiosarcomas [4]. Each type is a different entity. PAB arises from non-radiated parenchyma of the breast; it has no known risk factors, and it usually arises in younger women in the third or fourth decade of life. Secondary angiosarcoma is more prevalent; it is associated with a previous history of radiotherapy for breast adenocarcinoma; this has a latency of five or more years and is most often found in older women [5].

Patients with breast angiosarcoma present with an unspecific palpable mass rapidly increasing in size. The mean size of these tumours is about 6 cm in diameter. Diagnosis and treatment delays may lead to progressively larger breast masses with bluish skin discolouration. Metastases of AB are known to be hematogenous rather than lymphatic, and they occur more frequently in bone, liver, and lungs [6].

Diagnosis of angiosarcoma can be demanding; mammograms related to PAB are usually non-specific. These tumours frequently can be missed, especially if low-grade; they may present with completely normal findings with minor observation of skin thickening. Visible masses may appear oval or irregular and exhibit circumscribed or indistinct margins; some patients may present with focal symmetry in conjunction with coarse calcification [7]. Ultrasound may be helpful in some patients, as they may present masses of mixed hyperechogenic and hypoechogenic character and high vascularity on Doppler [8]. Magnetic resonance imaging (MRI) can be helpful to detect angiosarcoma according to the level of progression. A low-grade angiosarcoma shows progressive enhancement, whereas high-grade angiosarcoma presents with rapid enhancement and washout with frequent visualization of large draining vessels [9]. Histopathological findings on fine needle aspiration can be non-specific as PAB can resemble various breast abnormalities; nevertheless, different biomarkers distinguish angiosarcoma from invasive breast carcinomas (ductal/lobular). The absence of cytokeratin and the presence of endothelial markers such as CD31 and CD34 confirm the diagnosis, as presented in our original case report [10]. CD31 is most specific for endothelial markers, whereas CD34 is the more sensitive. Different grades of PAB have been described: grade one is angiosarcoma with inter-anastomosing vascular channel and subtle endothelial atypia with few mitotic figures; grade two tumour cells reveal moderate cell atypia and multilayering endothelial cells; grade three tumours show marked nuclear polymorphism, numerous mitoses, and necrosis [11].

Radical mastectomy is the primary treatment of angiosarcoma; lymph node dissection is arguably not helpful as the tumours are less likely to spread via the lymphatic system. Chemotherapy has been proven to reduce the rate of recurrence and increase survival [12]. However, little data exists regarding adjuvant radiotherapy in treating PAB. The prognosis of primary angiosarcoma of the breast depends on the size of the tumour, metastasis to other organs, and histological characteristics. Grade 1 angiosarcoma has five years survival of 76%, but this drops to 15% in grade 3 tumours [13].

The patient who presented in this case report was in her 50s, which is a distinctive presentation for a PAB. In our case report, the patient had immediate reconstructive surgery with a latissimus dorsi flap, which can be safely performed to cover a wide area exposed after angiosarcoma resection [14]. Unfortunately, she was lost to follow-up.

Conclusion

Primary angiosarcoma of the breast is a rare entity; it presents in a small percentage of breast masses. Diagnosing and identifying it could be challenging with its ambiguous mammography findings and resemblance to benign tumour presentation. It is crucial to report such cases in order to facilitate angiosarcoma as differential diagnosis and prevent rapidly progression of such tumours.

References

1. Lim RF, Goei R. Best cases from the AFIP: Angiosarcoma of the breast. Radiographics. 2007;27 Suppl 1:S125-S130.
2. Arora TK, Terracina KP, Soong J, Idowu MO, Takabe K. Primary and secondary angiosarcoma of the breast. Gland Surg. 2014;3(1):28-34.
3. Desbiens C, Hogue JC, Lévesque Y. Primary breast angiosarcoma: Avoiding a common trap. Case Rep Oncol Med. 2011;2011:517047.
4. Tomich J, Grove Nigro K, Barr RG. Primary angiosarcoma of the breast: A case report and review of the literature. Ultrasound Q. 2017;33(1):46-48.
5. Ohta M, Tokuda Y, Kuge S, et al. A case of angiosarcoma of the breast. Jpn J Clin Oncol. 1997;27(2):91-94.
6. Glazebrook KN, Magut MJ, Reynolds C. Angiosarcoma of the breast. AJR Am J Roentgenol. 2008;190(2):533-538.
7. Dashevsky BZ, Charnoff-Katz K, Shin SJ, Babagbemi K, Rosenblatt R. A case of primary breast angiosarcoma. Radiol Case Rep. 2015;8(3):741.
8. Varghese B, Deshpande P, Dixit S, Koppiker CB, Jalnapurkar N. Primary angiosarcoma of the breast: A case report. J Radiol Case Rep. 2019;13(2):15-25.
9. Pandey M, Martin MG. Primary angiosarcoma of the breast: A case report and review of literature. World J Oncol. 2014;5(3):144-148.
10. Taghipour Zahir S, Sefidrokh Sharahjin N, Rahmani K. Primary breast angiosarcoma: Pathological and radiological diagnosis. Malays J Med Sci. 2014;21(5):66-70.
11. Kim YS, Kim YJ, Yim KI, Park WC. A Case Report of Primary Breast Angiosarcoma with Fatal Pulmonary Hemorrhage Due to Thrombocytopenia. J Korean Surg Soc. 2012;82(4):251-255.
12. Cohen-Hallaleh RB, Smith HG, Smith RC, et al. Radiation induced angiosarcoma of the breast: Outcomes from a retrospective case series. Clin Sarcoma Res. 2017;7:15.
13. Rosen PP, Kimmel M, Ernsberger D. Mammary angiosarcoma. The prognostic significance of tumor differentiation. Cancer. 1988;62(10):2145-2151.
14. Mella JR, Ross KM, Li GZ, Pomahac B, Raut CP, Orgill DP. Cutaneous breast radiation-associated angiosarcoma: Anterior chest wall reconstruction options following extra-radical resection. Plast Reconstr Surg Glob Open. 2018;6(9):e1938.