

# Metastatic breast angiosarcoma: A case report

Clare Desira, Malcolm Buhagiar

## ABSTRACT

**Introduction:** Angiosarcoma of the breast is a rare finding that accounts for less than 1% of breast malignancies and less than 5% of all sarcomas. It is categorized according to the etiology: de novo (primary) and therapy related (secondary).

**Case Report:** This case report discusses the management of a patient (a 62-year-old female) who had primary breast angiosarcoma. Management of such a patient always merits a multidisciplinary team approach that involves the breast surgeons, oncologists, breast radiologists, and breast care nurses.

**Conclusion:** The main conclusion from this case report is that frequent breast screening would be beneficial in order to diagnose malignancy at an early stage, especially in patients who have a family history as survival depends on the stage at which the disease is diagnosed. However, this disease has a very poor prognosis overall.

**Keywords:** Breast angiosarcoma, Mammography, Metastasis, Oncology

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## INTRODUCTION

Breast angiosarcoma is a malignancy that arises from endothelial cells that line blood vessels supplying the breast. The prognosis of this rare malignancy is related to the grade at the time of diagnosis, and radical mastectomy remains the primary mode of treatment. It is typically aggressive with a high metastatic potential; however, it is unlikely to invade adjacent lymph nodes; instead it frequently metastasizes to bone, lungs, liver, and the contralateral breast. Diagnosis is made using biopsies, mammogram, and ultrasound. In young patients with denser breast tissue, magnetic resonance imaging (MRI) of the breast is preferred. A computed tomography (CT) thorax abdomen and pelvis would then be needed to determine the extent of disease spread and to plan first line management.

## CASE REPORT

A 62-year-old female presented privately to a breast surgeon in view of bilateral palpable breast lumps. A mammogram and ultrasound were performed. The mammogram showed scattered fibroglandular elements bilaterally, a well-circumscribed lesion with partially complete radiolucent halo in the upper inner left breast measuring 3.1 cm × 1.9 cm × 3.3 cm (Figure 1) which was deemed to benign, together with a mass in the medial right breast measuring 5.2 cm × 3.6 cm (Figure 2). The ultrasound further confirmed these findings, with no pathological axillary lymph nodes. The patient was therefore referred for a biopsy of the right breast lump.

An ultrasound (US)-guided biopsy of the right-sided mass was performed, and histology showed a vascular spindle cell neoplasm with a differential of a spindle cell hemangioma, with a suggestion for a wide local excision for a more definitive diagnosis.

This was performed successfully and the histology indicated a tumor of variable morphology. The diagnosis was that of intermediate grade angiosarcoma, the excision

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of which appeared to be complete. However, considering the nature of the malignancy, a right mastectomy was also performed. The tissue was sent for further histological analysis that excluded further foci of disease.

A CT of thorax, abdomen, and pelvis (TAP) was also performed, and this excluded distant metastases.

The patient was later reviewed by the surgical team post-operatively and she noticed that the left breast lump had grown. There were no overlying skin changes and no nipple discharge. The lump was mobile and was approximately 5 × 4 cm in size. She expressed her desire to undergo reconstruction of the right breast followed by a mastectomy on the left.

A US-guided biopsy of the left breast mass was done. There was no evidence of any axillary lymphadenopathy; however, it had shown an increased growth measuring 4.2 cm × 4.2 cm × 2.2 cm. Histology showed a benign/low grade spindle cell neoplasm which could represent a stromal component of a phyllodes tumor. A left mastectomy was therefore performed, and histology later indicated an encapsulated papillary carcinoma, foci of high grade ductal carcinoma in situ (DCIS), with two foci of metastatic angiosarcoma involving the skin.

This case was then discussed at the breast multi-disciplinary team (MDT) meeting and the patient was referred to oncology.

The new skin findings implied metastatic disease. Another CT TAP was performed, and this now showed multiple nodules in the subcutaneous fat, measuring up to 2.6 cm (Figure 3). A nodule over the left iliac bone was biopsied and was in keeping with metastatic angiosarcoma. An MRI thorax excluded the presence of recurrent/residual disease.



Figure 1: Mammogram showing lesion in left breast.

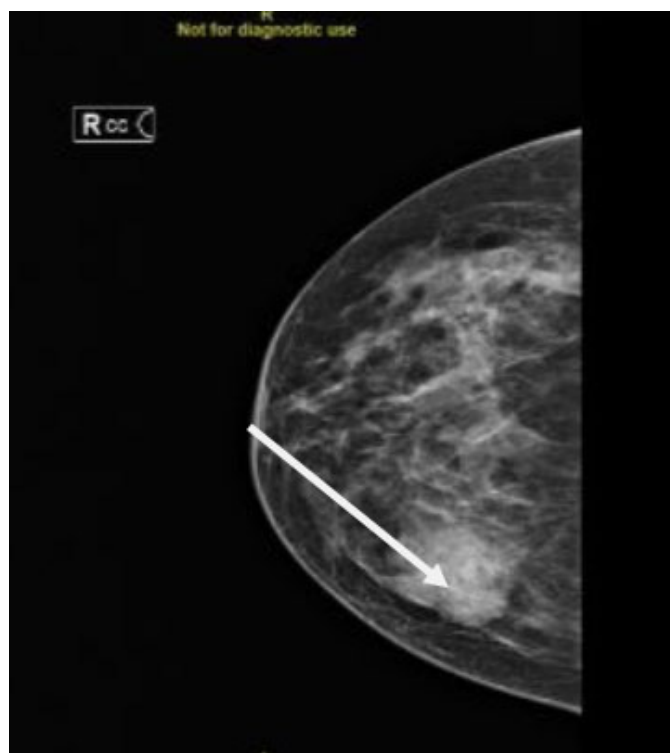


Figure 2: Mammogram showing lesion in right breast.



Figure 3: CT TAP image showing a subcutaneous nodule.

This patient was initiated on a course of liposomal doxorubicin from the oncologist with the intent of controlling disease and symptoms. She had a partial response to treatment, although her disease unfortunately progressed within a few months of treatment cessation. She is currently on pazopanib, a tyrosine kinase inhibitor.

Since she had a strong family history of malignancy, with multiple family members involved including both her mother and maternal aunt also having breast cancer, she was also referred for genetic studies.

## DISCUSSION

Breast angiosarcoma is defined as malignant proliferation showing endothelial differentiation. It is a rare tumor, accounting for around 0.04–0.1% of all breast malignancies. It is divided into two categories: primary, a vascular neoplasm that occurs in the breast parenchyma; and secondary that develops in the skin, chest wall, or breast parenchyma secondary to surgery (due to chronic lymphoedema after lymphadenectomy or mastectomy) or radiation therapy. Radiation-induced angiosarcomas are defined as sarcomas that occur in patients with a history of radiotherapy at least three years prior to the occurrence of the disease [1]. Although primary and secondary breast angiosarcomas share many similarities, there is evidence that the etiology and the natural history of the diseases might differ. Primary disease is typically found more frequently in younger women without a history of mammary carcinoma, while secondary disease is more commonly found in older women as a complication of the procedures mentioned above [2, 3]. The types of biopsies that need to be taken also vary between the two types. In primary a breast biopsy is needed to define the tumor, while in secondary a punch biopsy of the skin is typically done. Secondary angiosarcomas have a worse prognosis than primary and are more prone to local recurrence and distant metastasis. Since this patient had no recent history of radiation or surgery, this was therefore by definition a case of primary breast angiosarcoma. The patient also had a strong family history of malignancy making genetic testing crucial in management [2, 3].

Patients with breast angiosarcoma commonly present with an enlarging palpable breast lump [4]. Skin involvement is quite common in primary angiosarcoma, turning it a blue-red color, while skin nodules are noticed at the site of radiation in secondary malignancy [5]. Some clinicopathological features can help determine the prognosis of the disease. These include tumor grade, size, spread, and excision margin status. Some studies indicate that an increase in age, higher grade, and regional spread of the tumor result in a worse prognosis [6].

Complete surgical excision is the main treatment for breast angiosarcoma patients. Total mastectomy with adequate surgical margins is typically performed. Due to hematogenous dissemination of the angiosarcoma, axillary lymph node dissection is being deemed unnecessary. Angiosarcoma develops over a much wider field of the breast and chest wall than expected, and so total mastectomy tends to reduce local recurrence and thus improve prognosis when compared to breast conservation surgery. In the above case the patient underwent wide local excision of the lump on the right

breast, followed by bilateral mastectomies. The findings of metastasis indicate that she would need further chemo or radiotherapy to try and control progression of disease. In fact, in high grade angiosarcomas chemotherapy is showing a better outcome. With local recurrence radiotherapy might also be indicated [1, 3, 7].

Studies are looking at systemic therapies for treatment of angiosarcomas. A 2018 study indicates that treatment with vascular endothelial growth factor (VEGF) inhibitors seem to be promising. Vascular endothelial growth factor inhibitors are targeted therapies aimed at molecular abnormalities and have even less side effects than chemotherapy [8]. Other treatments such as immunotherapy, targeting interleukin-2, and even hyperthermia (using heat against malignant cells) are being studied in clinical trials [9].

## CONCLUSION

This patient presented with palpable breast lumps and was initially kept under surveillance. Delaying intervention for so long after detecting a breast lump is less than ideal, since earlier diagnosis of breast angiosarcomas enables better prognosis due to earlier management, where aggressive resection favors increased survival. Mastectomy with adequate tumor margins is recommended to reduce recurrence, along with close long-term follow-up. Frequent mammograms as part of the national screening program can help detect malignancy earlier, especially in patients with a known family history of malignancy.

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**Author Contributions**

Clare Desira – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Malcolm Buhagiar – Conception of the work, Design of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Authors declare no conflict of interest.

**Data Availability**

All relevant data are within the paper and its Supporting Information files.

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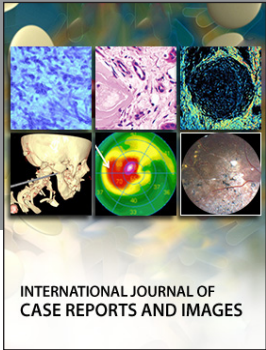
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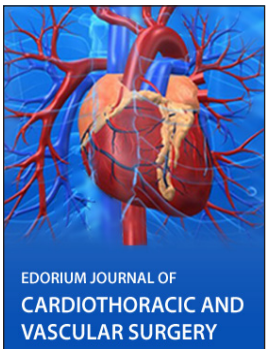
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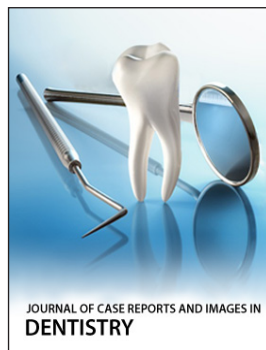
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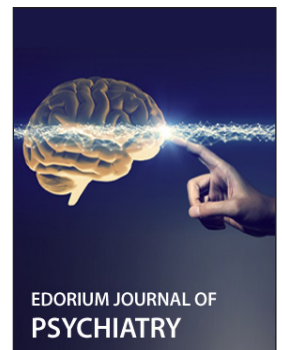
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