

Primary angiosarcoma of the breast: A case report and review of the literature

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Introduction

Angiosarcomas are uncommon malignant neoplasms characterized by extensively infiltrating anaplastic cells derived from blood vessels and lining irregular blood filled spaces (sinusoids). Primary angiosarcoma of the breast is a rare aggressive tumour of unknown etiology and may be difficult to diagnose. We present a case to illustrate the difficulty in diagnosis and the effect of this on the prognosis of the disease.

Case report

A 22 year old student presented at the surgical outpatient unit of LAUTECH Teaching Hospital, Osogbo, Nigeria with a right breast mass of 8 months duration. The swelling slowly but progressively increased in size with no nipple discharge. An excision biopsy of the mass done 9 months earlier in another centre suggested capillary haemangioma. Examination revealed a young lady apparently looking healthy. The left breast was normal but the right showed a circum-areola scar with a purplish lesion in the inferior and lateral aspects of the breast. There was a satellite nodule on the edge of the previous scar (Fig.1). An impression of recurrent haemangioma was made.

Breast ultrasonography revealed multiple hypoechoic solid masses of various sizes in the subcutaneous region of the areola and the outer quadrant. Colour flow Doppler study showed multiple vessels within and surrounding the masses but with no detectable increase of blood flow when compared to the normal left breast. Fine needle aspiration for cytology report suggested recurrent haemangioma. Excision biopsy with nipple preservation was repeated; histology report still came as hemangioma.



Figure 2: Presentation of the patient following the second excision

She reported back with recurrence and profuse bleeding 6 months after this and had an emergency excision with parascapular fasciocutaneous flap and skin grafting of the defect. Histology then showed grade 1 angiosarcoma.

She developed left hemiparesis ten days post operatively and had a brain computerized tomography scan which revealed multiple brain metastases to the cerebrum (Fig. 2). She was commenced on tablet of Dexamethasone 8mg three times daily while being worked up for palliative chemotherapy. She died three days later following a generalized tonic clonic seizure.



Figure 3: Left hemispheric metastasis

Discussion

Angiosarcoma are rare breast tumours. They represent a heterogeneous group of malignant vascular tumors occurring in different anatomic sites. In the female breast, they account for less than 1% of all malignant tumors and mainly develop as secondary angiosarcomas after prior irradiation¹.

Primary angiosarcoma of the breast is very rare²

though many cases of angiosarcomas following mastectomy and irradiation have been reported^{3,4,5}. This should be differentiated from Stewart-Treves syndrome which is a lymphoedema-associated angiosarcomas and usually developing in female patients after mastectomy and axillary lymph node dissection⁶. Studies have shown that angiosarcoma of the female breast represents a genetically heterogeneous tumour entity without a readily identifiable pattern of common chromosomal alterations⁷.

A frequent clinical presentation is a painful palpable smooth mass or diffuse enlargement of the breast without a palpable mass but with purple discoloration of the overlying skin. They are aggressive and tend to recur locally, spread widely, with lymph node and systemic metastases associated with a high rate of tumour related deaths⁸. These features are consistent with the clinical history of the patient in the present report.

Difficulty in making a diagnosis in angiosarcomas of the breast have been reported^{2, 5, 6} and the need for an extreme caution in the interpretation of histological characteristics of all palpable vascular tumours of the breast is emphasised⁹. Difficulty in making a definitive diagnosis based on cytological examination alone has been stressed; and that histology, supported by immunohisto-chemical studies, helps in arriving at the diagnosis¹⁰.

All angiosarcomas tend to be aggressive and are often multicentric. Treatment is therefore difficult. Complete surgical excision is mandatory. Even then, prognosis is poor¹¹ as exemplified by this case. Treatment is now standardized with radical mastectomy associated with adjuvant chemotherapy especially paclitaxel and docetax^{3, 4} and radiotherapy for grade 2 or poorly differentiated tumours². Angiosarcomas often recur after surgery; chemotherapy, radiotherapy, and targeted therapy against tumour biological properties¹²

may be a new approach to treatment. Among

sarcomas, angiosarcoma was the only histologic type significantly associated with a poorer outcome¹³ in a multivariate analysis¹³ with a ten year overall survival rate of 80% in low grade tumours and 20% for high grade tumours².

The need for extreme caution in the interpretation of the histological characteristics of all palpable vascular tumours of the breast is emphasised⁹. Vascular lesions of the breast must be thoroughly investigated before they can be described as benign. This will help in early diagnosis and reduce the mortality that accompanies angiosarcoma of the breast.

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