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Pathology

Primary Angiosarcoma of Breast in a Young Female- A Case Report

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Abstract **Case Report**

Introduction: Primary angiosarcoma of the breast is exceedingly rare, and represents around 0.04% of malignant breast neoplasm. Radiographically, breast angiosarcomas exhibit no pathognomonic features. They often appear as ill-defined masses on mammograms. The diagnosis is essentially based on the histological assessment of the excised sample. The treatment is based on simple mastectomy. This neoplasm carries a very poor prognosis, with a five- year survival of 8–50%. ***Case Report:*** We report a case of a 20- year-old woman with a highly vascular mass in her left breast which is suggestive of malignancy at radiology. The patient underwent a mastectomy. The histology of tumour showed dilated vascular spaces lined by atpical endothelial cells infiltratimg throughout the breast parenchyma. There are solid areas comprising highly atypical spindle or epitheloid cells with hyperchromatic round, oval or spindle nuclei with marked anisonucleosis. On immunohistochemistry the tumour was found positive for CD31, the diagnosis of Angiosarcoma Grade III was made. The patient is now receiving chemotherapy as known on two-month follow-up of the patient. ***Conclusion:*** Young women with solid-appearing breast tumors that are highly vascular at the time of biopsy should be considered malignant until proven otherwise.

Keywords: Young female, primary breast angiosarcoma, malignant breast neoplasm, CD31+.

INTRODUTION

Angiosarcoma of the breast is an exceedingly rare disease that may occur as a primary neoplasm or as a complication of radiation therapy after breast conservation [1]. The frequency of this rare tumor is 0.04% of primary mammary tumors and approximately 8% of mammary sarcomas [2, 3]. They are characterized by a high malignity and a fast growth rate and evolve to rapid local recurrence and the appearance of visceral metastases [4]. The diagnosis is essentially based on the histological assessment of the excised sample. The treatment is based on simple mastectomy [5]. This neoplasm carries a very poor prognosis, with a five-year survival of 8–50% [6].

CASE REPORT

The patient was a 20-year-old, unmarried, nulliparous woman with a chief complaint of left breast enlargement of nine months duration.

On physical examination, a hard, contracted-muscle mass was located in the left breast which measured 14* 12 cm and was accompanied by ulceration and necrotic changes in the overlying skin. No axillary lymphadenopathy was palpated. Tumor markers such as CEA, CA15-3, fetoprotein and CA19-9 were all within normal limits. In hormonal studies, luteinizing hormone (LH) and follicle-stimulating hormone (FSH) were within normal limits.

An ultrasound showed a diffuse and ill delimited hyperechogenic infiltration of the left breast which is hypervascular on doppler sonography. Mammography showed a non-specific and diffuses density area of about 12 cm. There was no microcalcification or distortion. The conclusion of radiologist was malignant finding (BIRADS4- 5).

A modified radical mastectomy was performed. The cut surface of the tumour appeared spongy and haemorrhagic measuring 12*9*5.5cm in size. Outer surface was covered by fibofatty tissue and was nodular. Cut surface showed a solid, red brown, well circumscribed mass measuring 7*6*4 cm. Microscopic examination of the mass demonstrated many dilated vascular spaces lined by atypical endothelial cells infiltrating throughout the breast parenchyma. There were solid areas comprising of highly atypical spindle or epitheloid cells with

hyperchromatic round , oval or spindle nuclei with marked anisonucleosis. There were 2-5 mitotic figures per high power field (hpf). Large area of hemorrhage & necrosis were presented suggestive of grade III angiosarcoma. On immunohistochemistry the tumour was positive for CD31. The patient is now receiving chemotherapy.

Figure 1: The cut surface of the resected specimen of the breast appears dark red and contains massive coagula

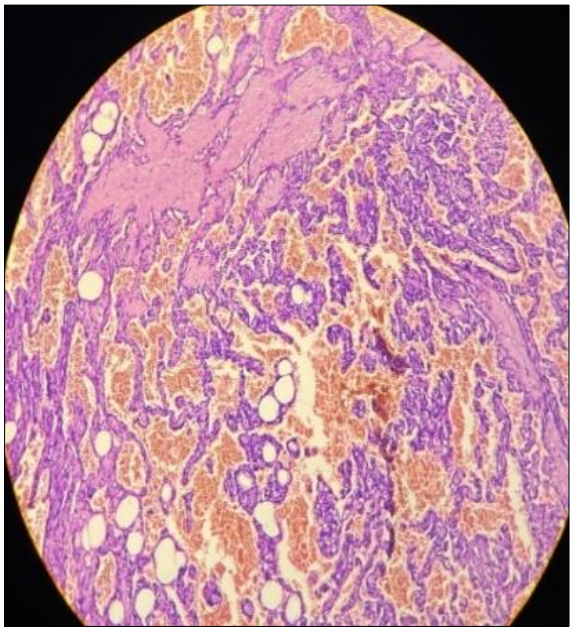

Figure 2: Histologic preparation shows that neoplastic endothelial cells demonstrate solid growth, occasional mitotic figures and sporadic protruding growth into the vascular lumen

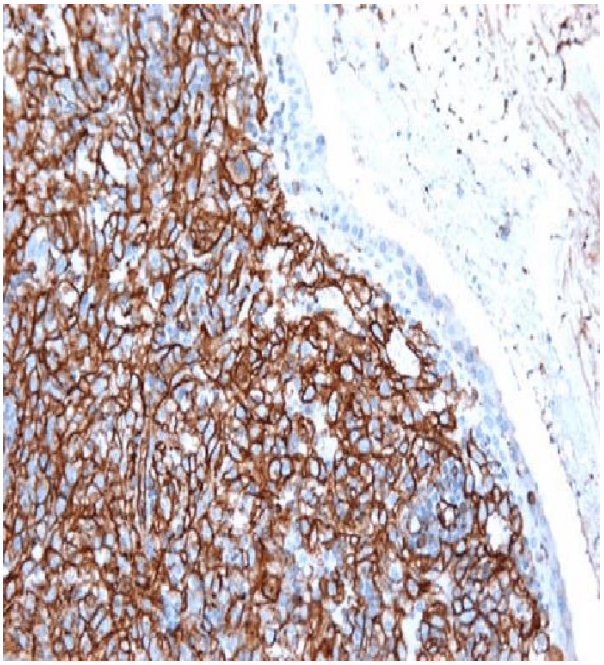

Figure 3: Immunoperoxidase staining for CD31shows the endothelial nature of the neoplastic cells

DISCUSSION

Mammary sarcomas are a heterogeneous group of malignant neoplasms that arise from the mammary stroma [7]. Angiosarcomas, one of the most common forms of mammary sarcoma, are developed from the endothelial lining of the blood vessels [8]. Breast angiosarcoma can be observed as a primary neoplasm or more commonly, is described in upper limb lymphedema as a result of mastectomy and radiotherapy for breast carcinoma [9]. Both primary and secondary breast angiosarcomas carry a prognosis worse than mammary carcinoma [10]. Synchronous bilateral angiosarcoma has been reported [10]. In the present paper, in accordance to the case reported, only primary angiosarcomas will be discussed.

Primary angiosarcoma of the breast is exceedingly rare, and represents around 0.04% of malignant breast neoplasms. Its incidence among breast sarcomas varies from 2.7% to 9.1%. [11]. Breast angiosarcoma is more frequent in young women (20 to 50 years) like in our case with no previous cancer history or other known risk factors [12, 13].

The right breast is more commonly involved than the left breast [14].

Angiosarcoma may have an insidious clinical onset, presenting as a painless often discrete palpable mass that grows rapidly [14, 15]. Some patients complain of a painful mass with tenderness. Approximately 2% of patients may present with diffuse enlargement of the breast. However, a bluish red discolouration of the overlying skin may be there [15].

Nipple retraction, discharge, or axillary node enlargement are generally absent. In most reported cases, the tumour size is >4 cm in diameter [16]. Most reports indicate that tumour size does not correlate with survival. The tumour size at discovery can reach 4 cm in diameter. Bilateral tumours have been reported and several cases have been diagnosed in the postmenopausal women.

Radiographically, breast angiosarcomas exhibit no pathognomonic features. They often appear as ill-defined masses on mammograms. Calcifications can be seen but differ from those seen with breast carcinomas [14].

Preoperative diagnosis, by FNAC and biopsy, may be difficult. Chen *et al.*, reported that the false negative rate of percutaneous biopsy was 37% [17]. Large-core biopsies might facilitate the correct diagnosis as they provide a larger sample [18], but such a macrobiopsy is often difficult to perform due to the vascular nature of these tumours.

Pathologically, these tumors are subdivided into three groups according to the classification proposed by Donnel and *et al.*, [19]. Grade I (well differentiated) contains open anastomosing vascular channels that proliferate within dermis, subcutaneous tissue or breast tissue. A single layer of endothelial cells lines these channels, which dissect through the stroma, causing distortion but little destruction of the preexisting lobules and ducts. The endothelial cells are usually flat; the nuclei may be hyperchromatic and contains small nucleoli. Solid and spindle cell foci, blood lakes, and necrosis are not present. Intermediate-grade angiosarcoma differs from low-grade by containing additional cellular foci of papillary formations and/or solid and spindle cell proliferation. The greater part of the tumor, however, is still composed of low- grade histology. Slightly increased mitotic activity is observed. In Rosen's study, intermediate-grade angiosarcomas behave more like low-grade sarcomas [20]. In grade III endothelial tufting and papillary formations are prominent. Conspicuous solid and spindle cell areas, mostly devoid of vascular formations, are present as well. Mitoses may be brisk, especially in more cellular areas. Areas of hemorrhage, known as “blood lakes,” and necrosis are also seen.

High grade angiosarcoma may contain low or intermediate grade elements, especially at the periphery of the tumor. These elements have deceptively benign appearing and are well differentiated [12].

Differential diagnosis of this rare tumor include: benign hemangioma, phyllodes sarcoma, stromal sarcoma, metaplastic carcinoma, fibrosarcoma, liposarcoma, squamous cell carcinoma with sarcomatoid features, myoepithelioma, fibromatosis, reactive spindle cell proliferative lesion, [16] and high-grade mammary carcinoma especially in small biopsy specimens containing only solid areas.

Immunohistochemical stains for endothelial cells show reactivity for several markers,including CD31, CD34 and von Willebrand factor (factorVIII). Among them, CD31 is considered the most sensitive and most specific endothelial cell marker and other sarcoma markers should help in making the correct diagnosis.

CONCLUSION

Young women with solid-appearing breast tumors that are highly vascular at the time of biopsy should be considered malignant until proven otherwise. Donnel's method for grading breast angiosarcoma is easy to implement and correlates well with clinical outcome. The treatment of choice is simple mastectomy. The role of radiotherapy and chemotherapy remains not fully established.

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