

A case report of hemangioendothelioma of the breast

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Citation

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Abstract

Vascular proliferations of the breast are uncommon but potentially diagnostically challenging lesions. Lesions with low grade malignant potential such as hemangiopericytoma and epithelioid hemangioendothelioma may rarely present in the breast. We report this case of hemangioendothelioma of low grade malignant potential in a 40 years old woman who presented with lump in the right breast since one year, gradually increasing in size. Excision biopsy revealed characteristic features consistent with hemangioendothelioma. We report this case in view of its rarity.

INTRODUCTION

The term hemangioendothelioma has become a useful designation for vascular tumors that have a biologic behavior intermediate between a hemangioma and a conventional angiosarcoma. Tumors included in this group have the ability to recur locally and have some ability to metastasize, but at a far reduced compared to angiosarcoma. Although the breast is an unusual location for this lesion, hemangioendothelioma should be considered a rare differential diagnosis of a breast mass.[1]

The frequency of this rare vascular tumor is 0.04% of primary mammary tumors and approximately 8% of mammary sarcomas. Several reports have been published with different names for this condition, such as hemangioblastoma, hemangiosarcoma and metastasizing angioma. [2]

CASE HISTORY

A female aged 40 years, presented with a painless lump in the upper outer quadrant of the right breast since 1 year. Fine Needle Aspiration (FNA) of this palpable right breast mass measuring 5X4cms was performed. Repeated FNA demonstrated hemorrhagic aspirate without any cellularity. Ultrasonography and mammography was advised to rule out highly vascular pathology.

Ultrasonography showed a well defined circumscribed hypoechoic lesion measuring 5.6X3.9cms in the right pectoralis muscle in the muscle plane in the axillary tail region.

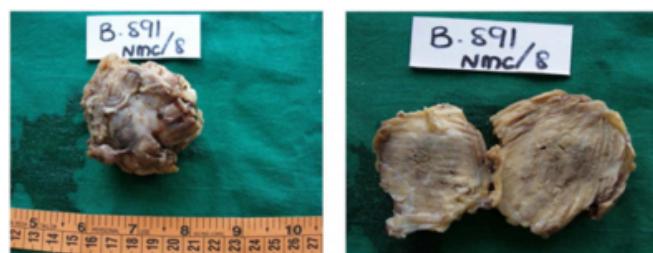
Doppler study revealed multiple tortuous hypoechoic areas suggestive of vessels which shows biphasic spectral wave form. A clinical diagnosis of soft tissue tumor with provisional diagnosis of vascular malformation was made.

The mass was excised and sent for histopathological examination.

MORPHOLOGICAL EXAMINATION

Grossly we received a single grey brown soft tissue mass measuring 5X4X4cms. Cut section was grey brown showing hemorrhagic areas with tiny cystic spaces.

Figure 1



Microscopic examination showed a circumscribed lesion composed of large round polygonal, slightly spindle shaped endothelial cells which are having vesicular nuclei and moderate amount of cytoplasm lining the vascular spaces. These cells were arranged in lobular pattern separated by fibrous septa. The lesion exhibited primitive vasoformation characterized by the presence of cytoplasmic vacuoles. Occasional mitotic figures were seen. There were no tumor giant cells, feeding vessels or areas of necrosis. In some foci the tumor cells lining the vascular spaces were seen

infiltrating into the adjacent muscle. The tumor cells reacted positively to CD 34 antigen

Figure 2

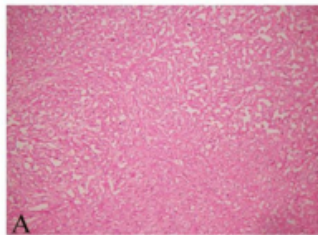


Fig.3 Section shows vascular spaces lined by large round polygonal, spindle shaped endothelial cells. H&E X100.

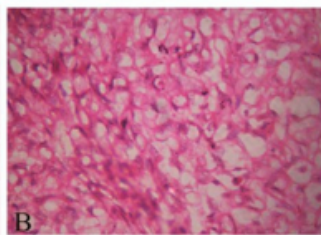


Fig.4 Endothelial cells showing cytoplasmic vacuoles.

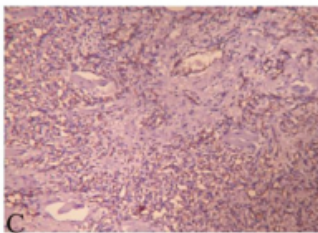


Fig.5 Endothelial cells showing CD34 positivity. CD34 X100.

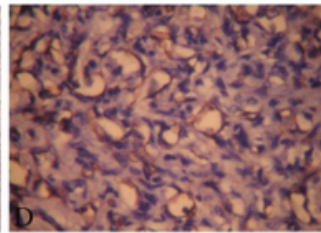


Fig.6 Endothelial cells showing CD 34 positivity. CD34X400

Based on the above histological findings, a diagnosis of hemangioendothelioma of low malignant potential was made.

DISCUSSIONS

Hemangioendothelioma is an uncommon vascular tumor with a predilection for pulmonary or hepatic involvement, but which may develop at almost any soft tissue site. This is an angiocentric vascular tumor, which can occur at any age . This is most commonly seen in the lower extremities, but rare cases in the breast have been reported.

Few cases have been found to be associated with polyurethane/ silicone breast implants.[3] Local recurrence and systemic metastasis may occur.

Histologically, the lesions most commonly have infiltrative margins and comprises of nests, cords and short strands of tumor cells within a myxohyaline stroma which may show varying degrees of degeneration.

Unlike the epithelioid hemangioma in which vascular differentiation proceeds through the formation of multicellular, canalized vascular channels, vascular differentiation in these tumors is more primitive and is expressed primarily at the cellular level. The tumors are

composed of short strands or solid nests of rounded to slightly spindled endothelial cells. The tumor cells form small intracellular lumen, which are seen as clear spaces, or “vacuoles” that distort or “blister” the cell. Frequently confused with the mucin vacuoles of adenocarcinoma, these miniature lumens occasionally contain erythrocytes. The stroma may show myxoid to hyaline change.

In most cases, the tumors appear quiet bland, and there is virtually no mitotic activity. In about ¼th of cases the tumors contain areas with significant atypia, mitotic activity (more than 1 mitosis per 10 high power fields), focal spindling of the cells, or necrosis. Such features can be correlated with a more aggressive course.

The immunohistochemical profile of this entity may initially suggest the diagnosis of metaplastic carcinoma, as epithelioid hemangioendotheliomas express cytokeratin CK 7 as well as CK18 in the majority of cases. However these tumors also express a wide range of vascular antigens (CD31, CD34 and factor VIII), not seen in breast carcinoma. In addition they are negative for high molecular weight cytokeratins, such as CK 14, whose presence is a characteristic feature of metaplastic breast carcinoma.[4]

A small percentage of benign appearing hemangioendotheliomas metastasize and cause the death of the patient. This occurs when the lesion has more atypical features. Because of low grade nature of these tumors, complete and ideally, wide local excision without adjacent radiotherapy or chemotherapy is the treatment of choice. Histologically malignant forms are treated similar to other sarcoma with atleast radical local excision.

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