CASE REPORT PEER REVIEWED | OPEN ACCESS

Epithelioid hemangioendothelioma arising from the brachial artery

Niharika Shah, Karki Smriti

ABSTRACT

Introduction: Weiss and Enzinger first described epithelioid hemangioendothelioma (EHE), a rare vascular tumor, in 1982 as a clinical state between hemangioma and angiosarcoma. Case Report: We present here a 40-year-old female with complains of pain and swelling over the right arm since five years. Intraoperative findings revealed a mass arising from the brachial artery. Histopathological examination, revealed, proliferation of chains and cords of epithelioid endothelial cells distributed in a myxohyaline stroma. The cells had eosinophilic cytoplasm containing vacuoles that deformed the cytoplasm. Some vacuoles contained fragmented erythrocytes. On immunohistochemistry, the tumor cells were positive for CD34. A final histopathological diagnosis of epithelioid hemangioendothelioma (low grade) was made. Conclusion: Epithelioid hemangioendotheliomas, especially primary vascular ones, are extremely rare, with 33 cases reported in English literature. Surgery seems to be the only beneficial treatment, with a doubtful role of chemotherapy and none for radiotherapy.

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INTRODUCTION

Epithelioid hemangioendothelioma (EHE) was first described in 1982 as a state purported to be between benign hemangioma and angiosarcoma by Weiss and Enzinger. It is a rare vascular tumor [1]. This led to the suggestion of a new term hemangioendothelioma owing to its intermediate clinical behavior between high grade angiosarcoma and benign hemangioma [2]. This type of tumor is rare, and is found in a range of ages, with presentation at varied sites in the body, and occasionally also metastasis, and multifocality. This lands one up with many differential diagnoses, further compounded by its bland morphology, with only subtle intracytoplasmic vascular lumina most of the times [1, 3].

Herein, we present a 40-year-old female with pain and swelling in the right arm since five years.

CASE REPORT

A 40-year-old female presented with complains of pain and swelling over the right arm since five years. There was no history of trauma. On examination the patient was hemodynamically stable, and there was a 2x3 cm

firm swelling, mobile, tender and non-pulsatile over the medial aspect of the right arm. Complete and differential blood counts and all other hematological parameters were within normal limits. A provisional diagnosis of a soft tissue tumor was made.

Ultrasonography of the right arm with Doppler study revealed a partly well defined oval hypoechoic lesion, within muscle at medial aspect of right mid arm surrounding and compressing the adjacent segment of brachial artery eccentrically with small branches from artery appearing to extend into the lesion, suggestive of organizing peri-vascular hematoma/ thrombosed pseudo-aneurysm/soft tissue tumor.

An excisional biopsy of the soft tissue mass in the right forearm with saphenous vein grafting was done. Intraoperative findings revealed a mass arising from the brachial artery present in the anterior aspect, approximately 5x2 cm in the inter-muscular plane adjacent to the brachial nerve obliterating the lumen. The excised tumor was sent for histopathological examination, and grossly, multiple irregular gray brown to focally blackish soft tissue measuring together 2x1x0.5 cm were received. Cut surface was solid gray white to gray brown.

Histopathological examination revealed proliferation of epithelioid endothelial cells in chains and cords against a myxohyaline stroma. The cells had intracytoplasmic vacuoles that deformed the eosinophilic cytoplasm with some even containing fragmented erythrocytes. These cells were mild to moderately pleomorphic with a vesicular to hyperchromatic nuclear chromatin, visible to prominent nucleoli. Several hemosiderin laden macrophages were also seen. No mitotic figures were identified, and the tumor cells were positive for CD34. A final histopathological diagnosis of epithelioid hemangioendothelioma (low grade) was made.

DISCUSSION

It is rather uncommon to encounter a primary vascular tumor. One might occasionally find a sarcoma or a leiomyosarcoma, which however tend to arise in large veins and not in small vessels or arteries. In our case, the mass was seen to be arising from the brachial artery obliterating its lumen.

The EHEs are a very rare occurrence within vessels, and these soft tissue tumors usually arise in the liver, lung, neck, bone or spleen [4]. There are only about 33 cases of EHEs reported in English literature, making these tumors extremely rare [5].

Microscopically, EHE lack well-formed vascular channels, with only subtle immature intracytoplasmic lumina present, occasionally containing fragmented erythrocytes and the cells are found in small nests, in single files or in cords. These tumors also lack a lobular pattern of growth as seen in benign hemangiomas and they infiltrate and encase surrounding structures.

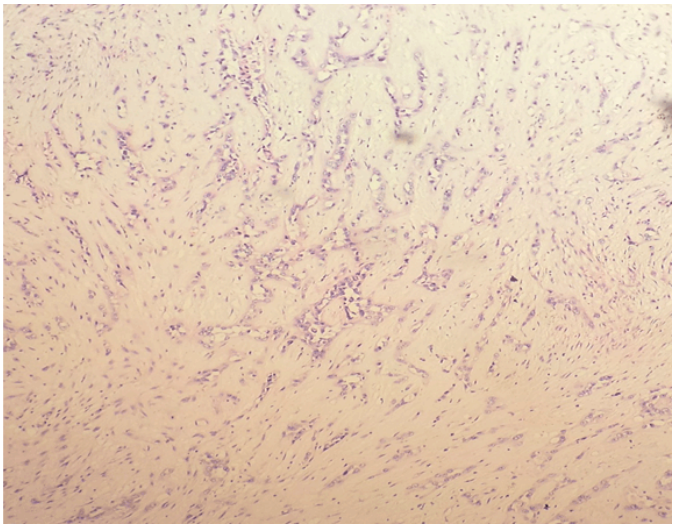

Figure 1: Proliferation of chains and cords of epithelioid endothelial cells distributed in a myxohyaline stroma (H&E, stain x100).

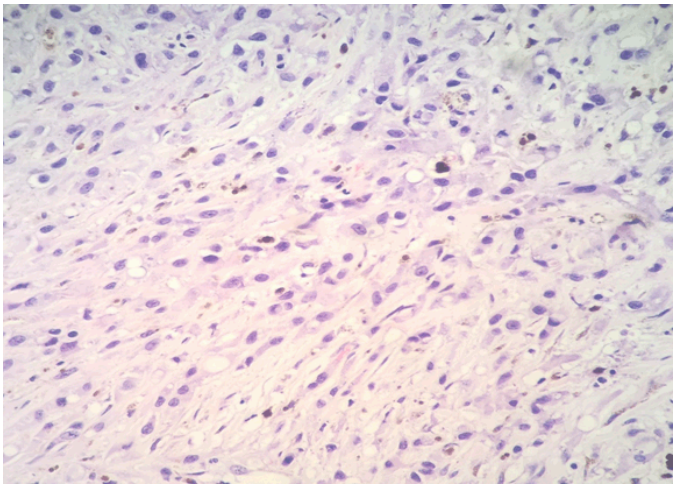

Figure 2: Tumor cells, with a vesicular to hyperchromatic nuclear chromatin and visible to prominent nucleoli, and an eosinophilic vacuolated cytoplasm. Occasional fragmented erythrocytes can also be seen within the cytoplasm (H&E stain, x200).

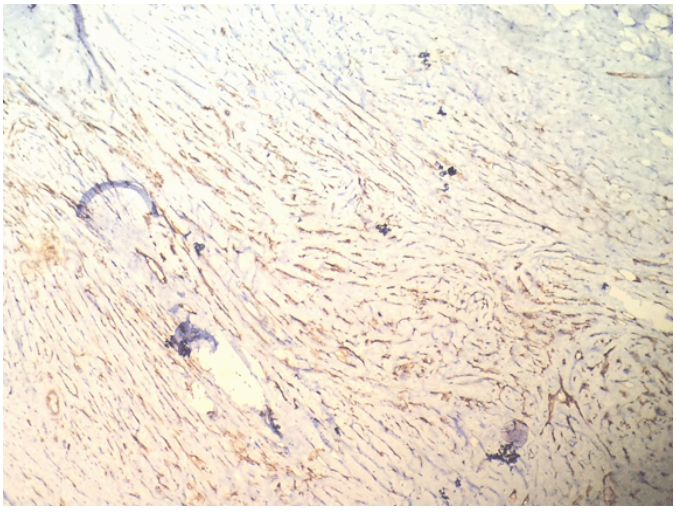

Figure 3: Tumor cells are positive for CD34 (magnification: x100).

The cells in EHE are distributed within a distinctive extracellular sulfated acid-rich matrix, which varies from a light blue (myxoid, myxo-chondroid), to a deep pink extracellular stroma. The cells, also known as blister cells, have intracytoplasmic vacuoles that deform a pale to more deeply eosinophilic cytoplasm [2].

Discovery of the nonrandom reciprocal t (1;3) (p36;q25) translocation [6], led to the detection of the corresponding fusion gene WWTR1-CAMTA1 [7, 8].

A family of calmodulin binding transcription activators, normally found only in brain, WWTR1, fuses to CAMTA1, on 1p36. Most cases of EHE examined so far, that reveal a classic morphology contain this translocation. Therapeutic available as of now have not been able to target it directly, however future studies may reveal downstream targets [2].

Stratification of this tumor into low and high risk groups has been attempted by utilizing a combination of mitotic activity and tumor size; patients with tumors 43 cm in diameter and having more than three mitoses/50 HPFs have a five-year disease-specific survival of 59% in contrast to 100% survival in patients whose tumors lacked these features [9, 10].

CONCLUSION

Epithelioid hemangioendothelioma treated via surgery have shown beneficial results with a doubtful role of chemotherapy and none whatsoever for radiation therapy. Lungs and lymph nodes are common sites of metastasis and are seen in an estimated 30% of patients and that too at a significantly later time.

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Author Contributions

Niharika Shah – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Karki Smriti – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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