Case Report

Epithelioid hemangioendothelioma: A rare vascular tumour

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ABSTRACT

Epithelioid hemangioendothelioma is a rare intermediate grade neoplasm of the vascular endothelium. This tumour has wide spectrum of behaviour having high rate of local recurrence, distant metastatic potential, and malignant transformation. We present a case of a 25-year-old female with epithelioid hemangioendothelioma of the left lower limb that was excised by wide local excision. There was no bone or skin involvement. The patient is on regular follow-up without recurrence since last one and a half years.

Key words: Epithelioid hemangioendothelioma, intermediate grade, vascular tumour

INTRODUCTION

Epithelioid hemangioendothelioma is an angiocentric vascular tumour with metastatic potential, histopathologically composed of epithelioid endothelial cells arranged in short cords and nests set in a distinctive myxohyaline stroma.[1] It is also known as intravascular bronchioloalveolar tumour, angioglomoid tumour, or myxoid angioblastomatosis. It is a very rare, intermediate grade vascular tumour that develops from peripheral veins of the lower limb, especially from the iliac and femoral veins.[2] It is important to recognize this neoplasm as it has wide spectrum of behaviour depending on the primary location of the tumour and surgical strategy differs like excision, large resection with bone reconstruction with or without irradiation or amputation.[3]

CASE REPORT

A 25-year-old female presented with painful progressive swelling in the right lower limb since 3 months. There was no history of trauma. Past and family history was not significant. On examination, an oval firm mass measuring 3 cm × 2 cm was palpable at the junction of middle and lower one-third of the right lower limb. The skin over the mass was unremarkable. There was no evidence of restriction of movement or paresthesia. X-ray of the lower limb revealed soft tissue opacity without bony involvement. Fine needle aspiration was attempted. Frank blood was aspirated. Smears were paucicellular showing occasional spindle-shaped cells against haemorrhagic background. Possibility of vascular neoplasm was considered. Excision biopsy was advised. On gross, the specimen was skin covered, measured 4.5 cm × 4 cm × 3 cm. Cut surface showed a 3.5 cm × 3 cm gray white firm mass surrounded by healthy tissue with grossly negative margins. Microscopic examination revealed the lesion arising from the vessel and extending into the adjacent soft tissue [Figure 1a]. Many small angulated lumina, delineated by flattened cells were seen [Figure 1b]. Tumour was composed of short strands, cords, and solid nests of rounded to slightly spindled eosinophilic endothelial cells. Occasional osteoclasts like giant cells are seen [Figure 2a]. Intracytoplasmic lumina containing erythrocytes were noted. Prominent clear vacuoles were seen within the cytoplasm of some epithelioid cells [Figure 2b]. Mitotic activity was not seen. The cells were embedded in a hyalinised stroma. Based on these histopathological features, a diagnosis of epithelioid hemangioendothelioma was made. Patient was advised regular follow-up and is disease free since last one and a half years.

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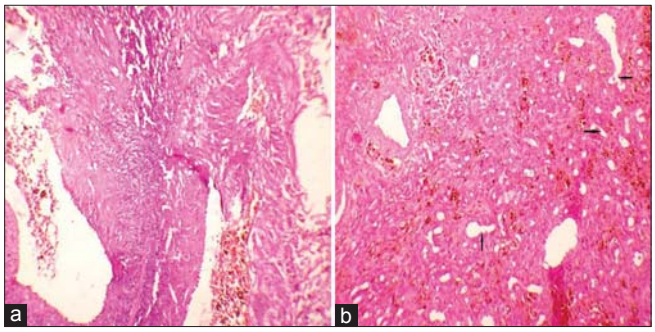

Figure 1: (a) Tumour arising from the vessel and extending into the adjacent soft tissue (H and E, ×100). (b) Many small angulated lumina (arrow) (H and E, ×200)

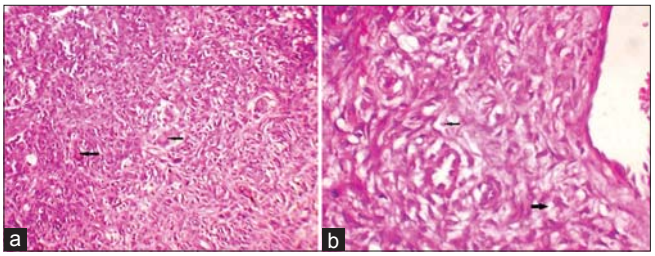

Figure 2: (a) Photomicrograph showing short strands, cords, and solid nests of rounded to slightly spindled eosinophilic endothelial cells along with giant cells (arrow) (H and E, ×100). (b) Intracytoplasmic lumina containing erythrocytes (thin arrow). Prominent clear cytoplasmic vacuoles in the epithelioid cells (thick arrow) (H and E, ×400)

DISCUSSION

Epithelioid hemangioendothelioma was described first in 1982 by Weiss and Enzinger[4] as a subgroup of the hemangioendotheliomas having an important endothelial or histiocytic component. They described 41 cases of epithelioid hemangioendothelioma in soft tissue. It has also been reported in liver, bone, gingiva, mediastinum, and lung.[5,6] The tumour develops as a solitary/rarely multicentric mass either in superficial or deep soft tissue of the extremities.[7] Nearly one-half to two-third cases originate from a vessel, usually a small vein. It occurs mainly in midadult life, affecting patients of either sex equally.

Microscopically, it is characterized by proliferation of rounded, eosinophilic epithelioid-like endothelial cells with frequent cytoplasmic vacuolization, a growth pattern potentially leading to a misdiagnosis of carcinoma. Spindle cells may also be seen, distinct primitive appearing vascular channels are formed, with erythrocytes occasionally seen in the lumina. The epithelioid-like endothelial cells exhibit features of normal endothelium, including positivity for keratin, CD 31, CD 34, factor VIII-related antigen and Ulex europaeus antigen.[7] The microscopic differential diagnosis includes epithelioid hemangioma, carcinoma, melanoma, epithelioid angiosarcoma, and epithelioid sarcoma. Cytogenetic involves identical translocation involving chromosomes 1 and 3 [t (1:3) (p36.6; q25)].

The biologic behaviour of these neoplasms differs depending on their anatomic positions and with regard to the age of occurrence. Mortality rate for epithelioid hemangioendothelioma of liver is 35% and lung is 65%. Prognosis in primary cutaneous lesion is good. The atypical histologic features suggestive of more aggressive behaviour are marked nuclear atypia, mitotic activity (>1/10 high power field HPF), spindling of the cells, and necrosis.[1,8] Based on studies which include both classic and malignant epithelioid hemangioendothelioma, the local recurrence rate in soft tissue lesions is 10-15%, metastatic rate 20-30%, and mortality 10-20%.[9] The present case did not reveal any atypical features. This case was of soft tissue origin without affecting the underlying bone, treated effectively by wide local excision with a disease-free interval of one and a half year and on a regular follow-up. Such tumours without atypical histologic features have a metastatic rate of 17% and mortality rate of 3%.[10]

CONCLUSION

Epithelioid hemangioendothelioma can mimic either benign or malignant tumours with unpredictable behaviour; thus, early diagnosis and treatment are particularly important. The case is presented due to its rarity, to highlight the importance of correct recognition of the entity and its biologic behaviour.

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