Epithelioid hemangioendothelioma: A rare primary vascular tumour in skin

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Abstract

Epithelioid haemangioendothelioma is a distinctive tumour arising from vascular epithelium characterized histologically by epithelioid endothelial cells arranged in strands or as individual units, in a myxoid or hyalinized stroma. This tumour is more commonly seen in deeper soft tissues and involvement of the skin is very rare. Only less than 10% of cases occur primarily in the skin. We are reporting a case presented to Teaching Hospital, Karapitiya with primary cutaneous epithelioid haemangioendothelioma involving the face. A 52-year-old female presented with a sclerotic skin plaque and nodules involving right side of the face for one month which developed spontaneous ulceration and bleeding. On examination revealed few violaceous firm nodules over forehead and right ear with sclerotic linear lesion over right cheek with ulceration at margin. No associated lymphadenopathy or systemic involvement. Computed tomography and MRI scan revealed ill defined mass in fatty tissue of skin in right cheek suggestive of skin malignancy without any internal involvement. Skin biopsy and immunohistochemistry confirmed the diagnosis of epithelioid haemangioendothelioma arising in skin and was managed with wide local excision of tumour.

Case history

A 52-year-old female presented to dermatology department, Teaching Hospital, Karapitiya with an asymptomatic slow growing skin plaque and few erythematous nodules involving right side of the face for one month. Spontaneous ulceration and bleeding were noted. No significant past history except recent onset weight loss and diffuse alopecia. An examination revealed few violaceous firm nodules over forehead and right ear with sclerotic linear lesion over right cheek with ulceration at margin (Figure 1). Associated lymphadenopathy, internal organ involvement or hepato spleenomegaly were not noted.

On investigation haematological, renal and hepatic parameters were normal. X-ray, computed tomography and MRI scan revealed ill defined mass in fatty tissue of skin in right cheek suggestive of skin malignancy without any internal involvement. Skin biopsy and immunohistochemistry from two different sites confirmed the diagnosis of epithelioid haemangioendothelioma arising in skin (Figure 2, 3).

She was treated with wide local excision of the tumour by onco-surgical team.

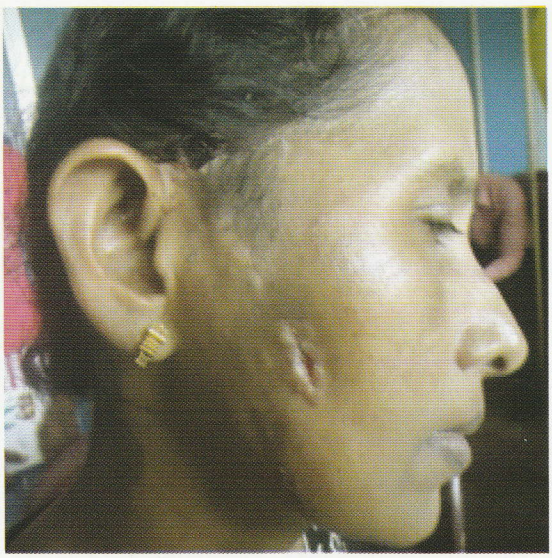

Figure 1.

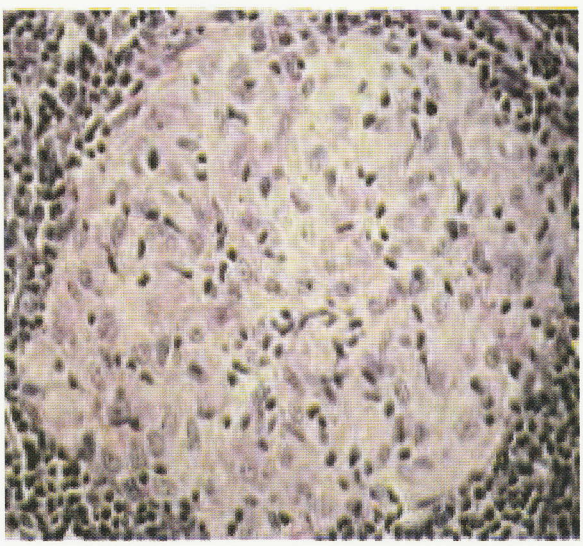

Figure 2.

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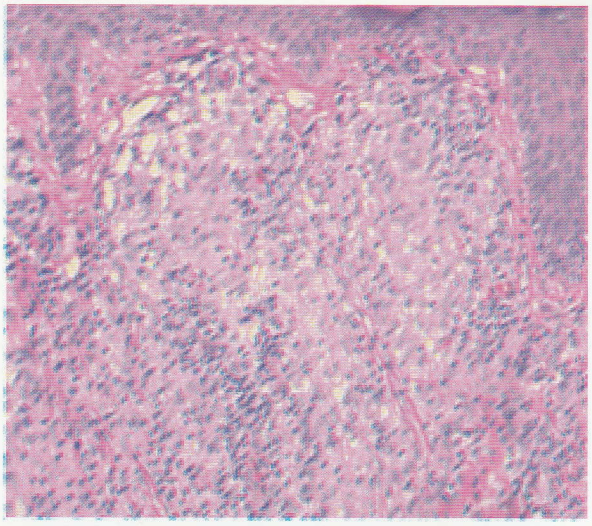

Figure 3.

Discussion

Epithelioid hemangioendothelioma is an angiocentric vascular tumour with metastatic potential[1,2]. There is no gender predilection. Cytogenetics involve identical translocation involving chromosome 1 and 3. Microscopically characterized by proliferation of rounded, eosinophilic epithelioid like endothelial cells with frequent cytoplasmic vacuolation. Spindle cells may also be seen. The epithelioid like cells show features of normal endothelium, including positivity for keratin, CD31, CD34. The biologic behavior of these tumours depend on their anatomic position and age of occurrence. Prognosis of primary cutaneous lesions are good. Atypical histology suggestive of more aggressive behavior are marked nuclear atypia, mitotic activity, spindling of cells, necrosis. Most tumors present as a solitary, slightly painful, soft tissue mass.

Skin involvement is often associated with an underlying soft tissue or bone tumor, but purely cutaneous tumours also occur. Overall, approximately 30% of patients develop metastatic disease involving regional nodes, lung, liver or bone and fewer than 50% of these patients with metastases have died of their disease. It is important to recognize this neoplasm early as it has wide spectrum of behaviour depending on the primary location of the tumour and surgical strategy differs accordingly.

References

1. Weiss SW, Bridge JA. Epithelioid hemangioendothelioma. In: Fletcher CD, Unni KK, Mertens F, editors. World Health Organization Classification of Tumours. Pathology and genetics of tumours of soft tissue and bone. 2 nd ed. Lyon: IARC Press; 2002; 173-4.
2. Gherman CD, Fodor D. Epithelioid hemangioendothelioma of the forearm with radius involvement: Case report. *Diagn Pathol* 2011; **6**: 120-4.
3. Castelli P, Caronno R, Piffaretti G, Tozzi M. Epithelioid hemangioendothelioma of the radial artery. *J Vasc Surg* 2005; **41**: 151-4.
4. Mentzel T, Beham A, Calonje E, *et al*. Epithelioid hemangioendothelioma of skin and soft tissues: clinicopathologic and immunohistochemical study of 30 cases. *Am J Surg Pathol* 1997; **21**: 363-74.