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CLINICAL RECORD

A rare angiosarcoma: retiform haemangioendothelioma

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Abstract

Objective: We report the case of a rare angiosarcoma, retiform haemangioendothelioma, in an 18-year-old young man, which presented as a recurrent ulcerating lesion of the left pinna.

Method: Case report and literature review of retiform haemangioendothelioma. This is a low grade angiosarcoma with a high local recurrence rate and low metastasis rate, and was first described in 1994 by Calonje *et al.*

Results: This patient represents only the third report of lymph node metastasis in a case of retiform haemangioendothelioma. To date, 31 cases of the tumour have been reported. Histological diagnosis of this group of vascular neoplasms can be challenging, as their histopathological appearance is intermediate between haemangioma and angiosarcoma.

Conclusion: Surgical excision remains the primary treatment modality, with adjuvant radiotherapy recommended in patients with large tumour size, local recurrence and lymph node metastasis, as seen in this case.

Key words: Ear; Pinna; Retiform Hemangioendothelioma; Hemangiosarcoma; Hemangioma; Neoplasm Metastasis

Introduction

Retiform haemangioendothelioma was first described only recently, in 1994.[1] It is a vascular neoplasm with a histopathological appearance intermediate between that of haemangioma and angiosarcoma, and thus represents a challenging, and potentially missed, diagnosis.

We present just such a misdiagnosed case of retiform haemangioendothelioma, previously managed as lymphangioma and treated with surgical excision. We describe the classical histological presentation of retiform haemangioendothelioma, as found in this case.

At presentation to our service, the patient had an extensive tumour, and subsequently required radical surgical excision and adjuvant radiotherapy, with associated cosmetic co-morbidity.

Case report

An 18-year-old young man presented with a one-year history of a discharging, ulcerated, thickened left pinna which had been repeatedly aggravated by rugby injuries (Figure 1). At the age of 11 years, he had been diagnosed with a congenital lymphangioma of the left side of the head and neck, which had been surgically removed but required further excision at the age of 13 years.

On examination, the patient had several areas of ulceration along the left pinna rim and post-auricular region.

Radiological examination of the head and neck identified a metabolically active mass involving the left pinna, suggestive of a vascular tumour, with signs of left mastoid osseous involvement (Figure 2). Enlarged level two and facial lymph nodes were also present.

Histological examination of a biopsy specimen indicated a vascular neoplasm.

Therefore, the patient underwent wide local excision, including total pinnectomy, with lateral temporal bone resection and neck dissection.

Macroscopic examination of the surgical specimen revealed a multifocal, tumourous mass 11 cm in maximum dimension. Histological examination showed numerous arborising, thin-walled, vascular channels in a retiform pattern, lined by plump endothelial cells, with a prominent lymphocytic infiltrate (Figures 3 and 4). Some areas showed a more solid growth pattern, with moderate but not severe atypia. A histological diagnosis of retiform haemangioendothelioma was made. Residual benign lymphangioma was also identified. A single, 2 mm metastatic deposit was found in a lymph node within the parotid tail.

The patient also underwent adjuvant radiotherapy. This, plus the radical nature of his tumour excision, resulted in moderate cosmetic co-morbidity.

Discussion

Retiform haemangioendothelioma was first described in 1994 as an extremely rare variant of low-grade angiosarcoma characterised by a high local recurrence rate and very low metastasis rate.[1] The majority of cases present on the trunk and limbs, with some isolated cases involving the head and penis,[2] although there is a female sex predilection.[1–3]

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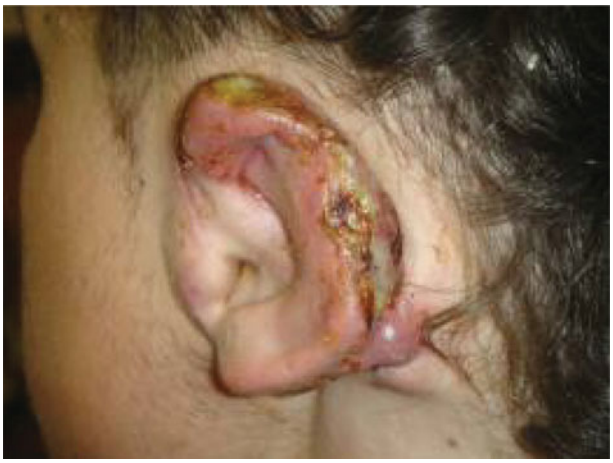

FIG. 1
The patient's left ear on presentation, showing the retiform haemangioendothelioma tumour.

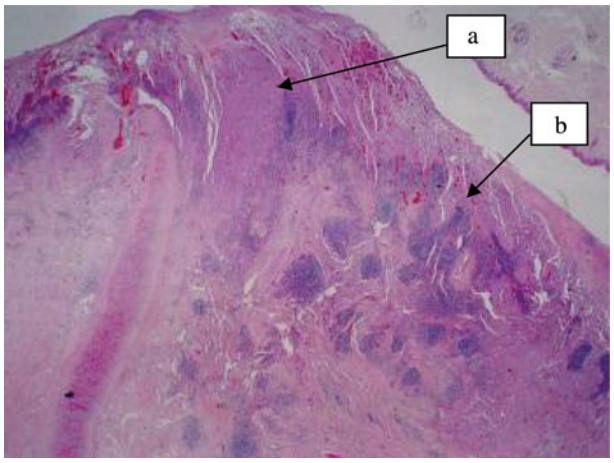

FIG. 3
Photomicrographs of the surgical specimen, showing (a) a solid area, and (b) a typical retiform area.

The tumour presents clinically as single, slowly enlarging, exophytic mass or a plaque-like tumour of the dermis and subcutaneous tissues.

There have been 31 previously reported cases of retiform haemangioendothelioma, and three cases (including the present case) of lymph node metastasis.[1,3] The existence of soft tissue metastasis remains controversial.[4] Our finding of a third case with lymph node metastasis highlights the need to continue to screen for metastasis, as the true frequency of metastatic spread cannot be definitively predicted from the current literature.

The diagnosis of retiform haemangioendothelioma is based on the presence of characteristic histological features. The differential diagnosis prior to biopsy includes lymphoma, haemangioma, dermatofibrosarcoma protuberans, cutaneous angiosarcoma, bacillary angiomatosis and Kaposi's sarcoma.[4,5] An association with previous lymphangioma or lymphoedema has been described. Retiform haemangioendothelioma shares some clinical characteristics with Kaposi's sarcoma, a tumour with known human herpes virus (HHV) 8 association. There has been a single reported case of retiform haemangioendothelioma in which HHV8 DNA sequences were detected, suggesting a potential viral aetiology.[6]

The term haemangioendothelioma refers to a group of vascular neoplasms that have a histopathological appearance intermediate between that of haemangioma and angiosarcoma.

In the present case, the histological appearance consisted of numerous elongated, arborising, thin-walled, vascular channels arranged in a retiform pattern, i.e. reminiscent of normal rete testis.[1] Some small areas of this tumour showed a more solid growth pattern (Figure 3), as first described in 1994 by Calonje *et al.* Other commonly seen histological features present in this case included a prominent lymphocytic infiltrate and papillae with hyaline collagenous cores (Figure 4), similar to a Dabska tumour. Cytological atypia is minimal and mitotic figures virtually absent.[7] Histological features associated with improved prognosis include the presence of differentiation toward the high endothelial cells of post capillary venules in retiform haemangioendothelioma.[1]

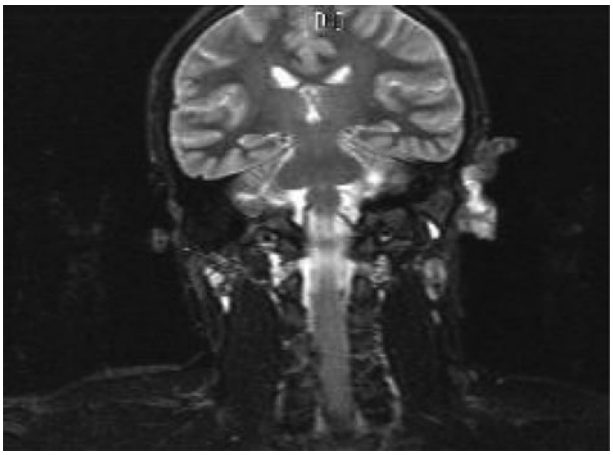

FIG. 2
Coronal computed tomography brain scan, showing a metabolically active mass involving the left pinna, suggestive of a vascular tumour, with signs of left mastoid osseous involvement.

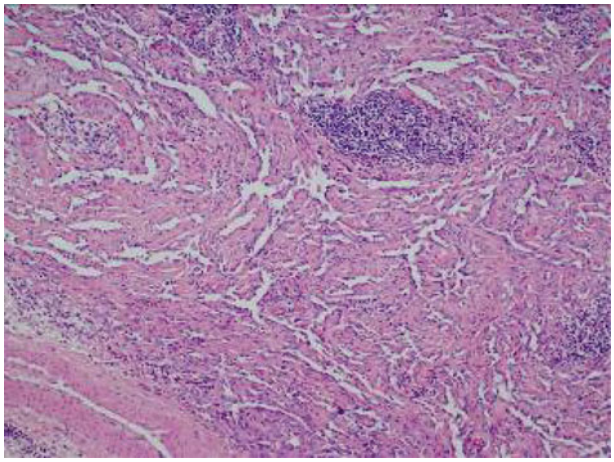

FIG. 4
Photomicrograph of the surgical specimen, showing retiform growth with lymphoid infiltrate.

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There have been no reported deaths associated with retiform haemangioendothelioma, although few cases have had long term follow up. The initial case series reported a median follow up of 7.35 years, with no mortality.[1–3,8]

The treatment of choice is surgical excision with histopathologically proven tumour-free margins. Since our patient's tumour was large and the margins close, and there was a single lymph node metastasis, he underwent adjuvant radiation therapy. This has previously been effective in cases with lymph node metastasis, large tumour size and local recurrence.[1,6]

Sixteen months post-operatively, our patient remained well. However, given his tumour's persistent recurrence, and the presence of a metastatic lymph node, the prognosis remained somewhat guarded.

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