Intravascular Epithelioid Haemangioma Of Temporal Artery:  
A Diagnostic Difficulty

PAI K, GUPTA A.

ABSTRACT

We report a case of Epithelioid haemangioma in a 23 year old female, arising in a branch of temporal artery presenting as a painless, pulsatile nodule in the temporal region, which was difficult to differentiate histologically from giant cell arteritis with thrombotic occlusion and neovascularisation. Laboratory investigations (Peripheral eosinophilia, and normal Erythrocyte Sedimentation rate and C-Reactive Protein) as well as clinical presentation helped in arriving at a diagnosis of Intravascular Epithelioid haemangioma.

Introduction

Epithelioid hemangioma (EH) is a benign vascular lesion that is characterized by well-formed, capillary-sized vessels lined by histiocytoid or epithelioid endothelial cells and often accompanied by a secondary inflammatory infiltrate. Whether or not these lesions are reactive or neoplastic has been debated and is reflected in the various designations, such as angiolymphoid hyperplasia with eosinophilia (ALHE), inflammatory angiomatous nodule, pseudopyogenic granuloma, atypical pyogenic granuloma, papular angioplasia, subcutaneous angioblastic lymphoid hyperplasia with eosinophilia and lymphofolliculosis, intravascular atypical vascular proliferation, histiocytoid haemangioma, and epithelioid haemangioma. pseudopyogenic granuloma[1] We report a rare case of epithelioid hemangioma arising from the temporal artery without history of trauma.

Case Summary

A rare case of epithelioid hemangioma arising in the right temporal artery of a 22-year-old woman was investigated. The lesion presented as a slow growing nodule over the temporal region since 6 months. The patient complained of pain over the nodule, but denied of headache, visual problem or any other complaints. On clinical examination, a small slightly pulsatile nodule over the temple measuring 0.5cms in diameter was felt. A clinical diagnosis of haemangioma /vascular lesion was made.

Table/Fig 1

Low power magnification shows an intravascular lesion, H&E, 100X
Intravascular lesion shows proliferation of blood vessels and diffuse lymphocytic infiltrate 400X, H&E

The histopathologic examination of the specimen showed cross-section of vessel, showing a sheet-like proliferation of the epithelioid endothelial cells in the lumen, which was almost occluded, with focal rupture of the media. There was diffuse inflammatory cell infiltration composed mainly of small lymphocytes along with scattered eosinophils and an occasional giant cell. A close histologic differential diagnosis was giant cell arteritis with neovascularisation secondary to organisation of thrombus in view of the presence of inflammatory cells and giant cell, though there was absence of granulomas. As the histologic differentiation was difficult,

Table/Fig 3

shows intravascular proliferation of numerous blood vessels; central lumen identified, Reticulin, 100X

C-Reactive protein (CRP), Erythrocyte Sedimentation rate (ESR) and Eosinophil count were requested. ESR and CRP were within normal limits. Peripheral eosinophilia of 13% was present. A diagnosis of Epithelioid Haemangioma-Intravascular was given taking into account the clinical, histologic and laboratory findings. Immunohistochemical stains could not be done due to resource constraints. The patient was reassured of the benign nature of the lesion. She has no complaints 7 months after excision of the lesion.

Discussion

Epithelioid Haemangioma is an unusual but distinctive vascular tumor that was first described by Wells and Whimster as Angiolymphoid hyperplasia with eosinophilia [2]. It presents as one or a few lesions on the head and neck of young adults, as a superficial lesion in the dermis or subcutis, but may sometimes involve deep soft tissue and in rare cases involve or arise in the blood vessel. Its pathogenesis is unknown, but probably represents a true vascular neoplasia or a reactive cicatricial phenomenon after a variety of aggressions, such as trauma, infection, or humoral disequilibrium.

In a study by Olsen and Helwig, more than 50% of the cases showed the presence of arterial structure, confirmed by the presence of an internal elastic lamina in close association with venular structures or was the site of endothelial cell proliferation [3]. Epithelioid Haemangioma shows characteristic histologic features, including a proliferation of small blood vessels, many of which are lined by enlarged endothelial cells with uniform ovoid nuclei and intracytoplasmic vacuoles. In addition, a perivascular and interstitial infiltrate composed primarily of lymphocytes and eosinophils is present. Eosinophils typically comprise 5-15% of the infiltrate [3]. Rarely, they can account for as much as 50% of the infiltrate. Occasionally, the infiltrate is devoid of eosinophils. Lymphoid aggregates with or without follicle formation are typical. Presence of giant cells is not reported in literature. Approximately 20% of patients have blood eosinophilia. The fact that there was no eosinophilia does not invalidate the diagnosis, since this characteristic is found in not more than 10-15% of the cases [3]. Eosinophil infiltrate in the present case constituted about 8% of the infiltrate.

A similar case of epithelioid hemangioma arising from the temporal artery without a history of trauma clinically mimicking temporal arteritis was
reported by Kitamura H et al [4]. Intravascular lesions of this tumor can cause diagnostic problem to the pathologist and such lesions have been described as Intravenous atypical vascular proliferation [5].

Based on the growth pattern of the epithelioid endothelial cells, John F et al grouped epithelioid haemangioma into "typical," and exuberant or "atypical." The latter examples had a prominent centrally located zone where nests or sheet-like aggregates of epithelioid endothelial cells did not form discrete vessels[6]. Our case showed exuberant proliferation of endothelial cells, without formation of vessels at places, and we regard it be a atypical growth pattern of epithelioid haemangioma.

There is frequent misdiagnosis with Kimura's disease. Although Kimura's disease was considered to be identical to angiolymphoid hyperplasia with eosinophilia (epithelioid haemangioma), studies suggest that they are different clinicopathological entities, bearing only superficial histologic similarities[7]. There is still however frequent misdiagnosis with Kimura's disease.

Optimal management appears to be complete local excision with periodic follow-up visits to monitor for local recurrence. Spontaneous remission may occur. No recurrence is noticed in our patient 7 months after excision.

**Conflict of Interest:** None declared

**References**


