Multiple cavernous haemangiomas of the the orbit and conjunctiva: A rare association

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Citation: Rizvi S, Yousuf S, Maheshwari V, Khan R. Multiple cavernous haemangiomas of the the orbit and conjunctiva: A rare association. JSCR. 2012 8:8

ABSTRACT

Cavernous haemangioma(CH) is mostly intraconal, single and unilateral in location. A 32 year old female presented with painless progressive growth near the inner canthus of right eye along with swelling of right upper eyelid and superomedial quadrant of the right orbit for five years. MRI showed lesions involving the preseptal space and extraconal compartment of the orbit at the superomedial aspect continous with the conjunctival swelling. Excision biopsy of the growths via anterior orbitotomy (vertical eyelid split technique) was done. Histopathological findings confirmed orbital cavernous haemangioma along with conjunctival CH.

We report a rare case of multiple cavernous haemangiomas arising from the conjunctiva as well as the superomedial orbit. Complete removal of the tumour is possible even in such difficult cases as the CH is totally encapsulated and meticulous surgical dissection can give good cosmetic result to the patient.

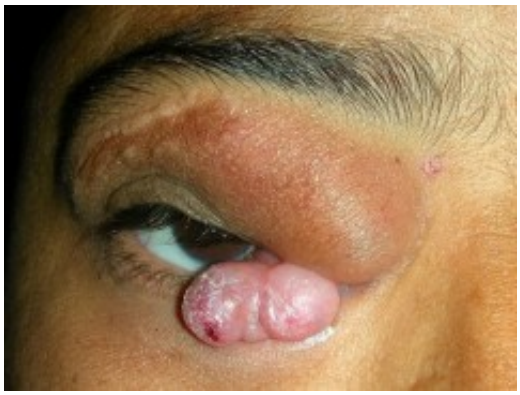
INTRODUCTION

Cavernous hemangioma (CH) is the most common benign non-infiltrative neoplasm of the orbit and is classified as a congenital low-flow vascular malformation. It results from formation of new vessels, proliferation of tissue components of the vessel wall and hyperplasia of cellular elements concerned with genesis of vascular tissue (1). Most of the cavernous hemangiomas are intraconal and lateral in location. It is typically single and unilateral (2). Cavernous hemangioma of the conjunctiva is rare and has been reported previously in only a few cases (3).

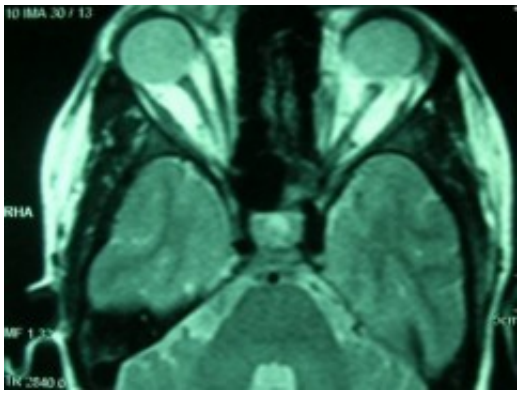
A rare case of multiple cavernous hemangiomas with an unusual extraconal and superomedial location and arising from conjunctiva is presented here.

CASE REPORT

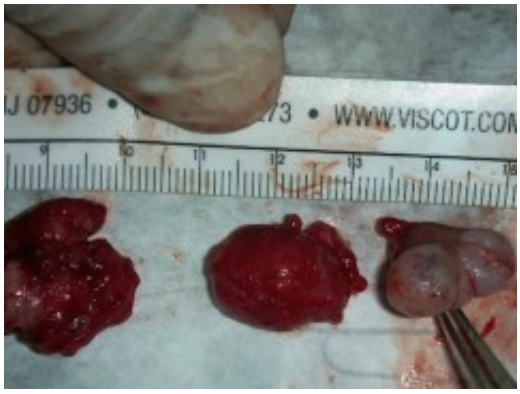
A 32yr old female presented with painless progressive growth from the bulbar conjunctiva near the inner canthus of the right eye along with swelling of the right upper eyelid and superomedial quadrant of the right orbit for five years. On ocular examination, the patient had mechanical ptosis. Visual acuity was 20/20 in both the eyes. Proptosis as well as other abnormality of any other ocular structure was not seen in the present case.



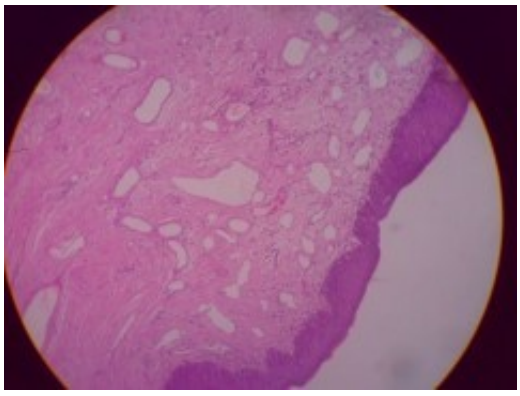
On local examination a smooth, oval, reddish in colour, bilobulated swelling of 15 x 17mm size arising from the bulbar conjunctiva near the inner canthus was seen. It was soft, vascular, compressible and freely mobile. Two swellings were felt in the right upper lid and superomedial quadrant of the right orbit respectively. The swellings were firm in consistency with well-defined margins, not attached to any underlying structures and the skin over it was freely mobile. The swelling did not change in size with valsalva manoeuvre, coughing, straining or change in head posture. (Fig. 1a)



MRI of the right orbit showed well defined altered signal intensity lesions involving the preseptal space and extraconal compartment of the orbit at superomedial aspect, which was continous with the conjunctival swelling. There was evidence of intense post contrast enhancement and a feeding artery extending up to the lesion.(Fig. 1b)



Excision biopsy of the growth via anterior orbitotomy with vertical eyelid split (Bryon Smith) technique was carried out. Three globular growths were dissected and excised. A bilobular conjunctival mass of 15×17mm, globular mass of 15×20mm and third lid swelling of 20×20mm was seen. (Fig. 2a)



Histopathology showed large blood filled spaces separated by fibrous stroma with flattened rete pegs of the overlying squamous epithelium, dilated and proliferating capillaries consistent with cavernous haemangioma (Fig. 2b). No recurrence of tumour was seen at 1 year of follow up.

DISCUSSION

Cavernous tumours are most common benign tumours of the orbit with a female preponderance. They are usually present in the second to fourth decades of life (4). Cavernous hemangioma mostly occurs as a well defined, encapsulated intraconal mass which is often unilateral and solitary and present as slowly progressive proptosis.

CT findings of CH typically demonstrates the presence of a well defined oval or round shaped mass but MRI provides superior evaluation of the orbit (5). MRI shows T1W hypo-isointensity and T2W hyperintensity. Variable homogenous or inhomogeneous contrast enhancement is the rule (6). These lesions have a small arterial input with small venous outflow channels and very slow flow within. These typical radiological features are seen in the present case.

Multiple cavernous hemangiomas of the orbit are extremely rare (7). Conjunctival cavernous hemangioma is also a rarity. It arises often from the conjunctival vessels and rarely from the scleral, muscular or orbital vessels (8) .We here present a case of multiple cavernous haemangioma of orbit, which was superomedial in location and associated with conjunctival hemangioma; a rare entity. There are very few case reports of multiple CH of the orbit. Nagulic *et al* reported excision of eight CH from a patient in a span of 24 years (2). Wang XG *et al* in their study found four cases of multiple CH out of 214 patients of orbital CH (7). In the present case three CH were excised out of which two were orbital CH and one was conjunctival CH.

Reports of conjunctival CH are much rarer still. Rao reports a case of conjunctival CH arising from bulbar conjunctiva near inner canthus (8). More recently another conjunctival CH arising from caruncular region over the bulbar conjunctiva was reported (3).The case reported by us was also arising from bulbar conjunctiva near inner canthus but its uniqueness lies in its bilobed appearance and its association with orbital CH.

A few authors also reported Extraconal CH. Rama et al reported a case of CH which was extraconal with superomedial location(9). Our case too had similar location but had multiple swellings.

One worker has reported concomitant appearance of orbital CH and venous angioma (10) but none of them has ever reported simultaneous occurrence of orbital and conjunctival CH.

Detailed search on MEDLINE could not find any such case with simultaneous occurrence of multiple extraconal orbital and conjunctival cavernous haemangioma reported earlier in the literature.

Although rare, cavernous hemangioma should be considered in the differential diagnosis of patients with multiple orbital mass lesions. Conjunctival CH usually are asymptomatic except for the cosmetic factor. Complete removal of the tumour is possible even when the CH is totally encapsulated and meticulous surgical dissection can give good cosmetic appearance to the patient.

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