Case Report

Intraorbital, extraconal cavernous hemangioma: a common tumor at an uncommon site-a case report

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Abstract

A wide variety of processes can produce space occupying lesions in and around the orbit including benign neoplasms, malignant neoplasms, vascular lesions, inflammatory diseases, congenital lesions etc. Cavernous hemangioma is the most common benign non-infiltrative neoplasm of the orbit. It is also the most common intraorbital tumor found in adults. Although histologically benign, they can encroach on intraorbital or adjacent structures and be considered anatomically or positionally malignant. We present here a case of extraconal, intraorbital cavernous hemangioma in a 21 years old male diagnosed histopathologically. So, we conclude that even though intraorbital cavernous hemangiomas present most frequently at intraconal locations, extraconal site for its occurrence cannot be ignored and must be considered in evaluation of intraorbital vascular lesions.

Key words: Cavernous hemangioma, Intraorbital, Extraconal, Vascular lesions.

Introduction

A wide variety of processes can produce space occupying lesions in and around the orbit including benign neoplasms, malignant neoplasms, vascular lesions, inflammatory diseases, congenital lesions etc [1]. Cavernous hemangioma is the most common benign non-infiltrative neoplasm of the orbit [2].The most frequent locations are retrobulbar muscle cone, especially lateral aspect of intraconal space, however, extraconal location is uncommon [1,2]. We present here a case of intraorbital cavernous hemangioma with extraconal location.

Case Report

A 21 years old male presented with progressive swelling of right eye and protrusion of the same eye forwards and laterally since five months. The swelling was not associated with burning sensation, blurring of vision, redness, doubling of vision or discharge from eye. The swelling did not increase with coughing, straining, valsalva maneuver, or change in head position. The patient was operated for a similar swelling 8 year back, histopathology report of which had shown the features suggestive of benign epithelial cyst.

MRI of orbit showed an ill-defined T1 isointense and STIR hyperintense lobulated lesion measuring 3.3x5.6x2.7cm in the extraconal portion of right orbit in the inferomedial quadrant. The mass is pushing the globe anteriorly with resultant proptosis of about 8cm as compared to the left side. The inferior and medial recti muscles are displaced superolaterally. Optic nerve is normal. Thus, a diagnosis of right introrbital, extraconal hamartoma was made.

Patient was operated with excision of the mass through endoscopic sinus surgery under general anaesthesia.

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Histopathology

Grossly, the excised specimen was a vaguely nodular mass measuring 4x3.5x3cm. The cut surface showed large grey brown hemorrhagic areas and focal grey white areas were also seen.

Figure 1: Gross picture of the specimen. (A) External surface.(B) Cut surface.

Microscopy from multiple sections of the specimen showed large, dilated, blood-filled vessels lined by flattened endothelium. The vessels were seen arranged in a roughly lobular arrangement or in a diffuse haphazard pattern. The walls were occasionally thickened and stroma showed mixed inflammatory cell infiltrates. Also seen were thrombi in various stages of organisation and recanalization. Thus, a diagnosis of ***'cavernous hemangioma'*** wasmade.

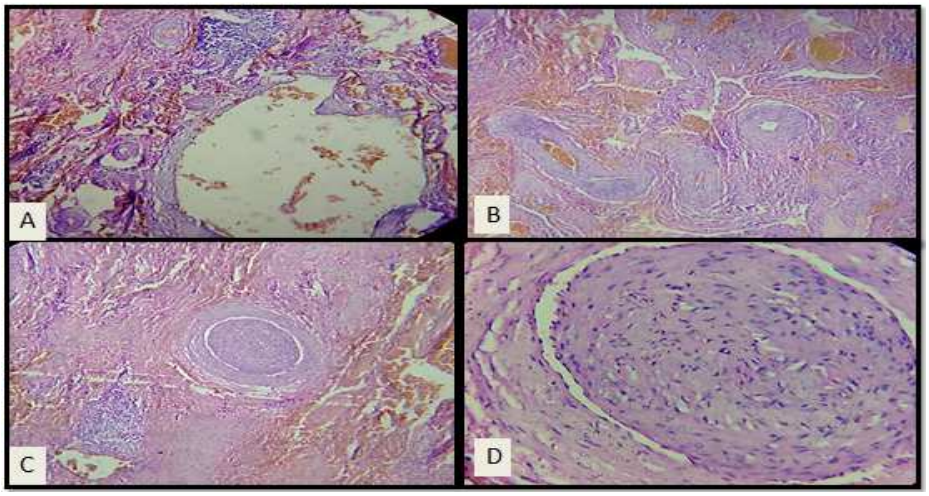

Figure 2 (A) and (B): large, dilated, blood-filled vessels lined by flattened endothelium (10x, H & E);(C) and (D): organising thrombus and recanalization. (10x, 40x, H&E)

Discussion

Hemangiomas occupy a gray zone between hamartomatous malformations and true neoplasms. They are frequently designated and regarded as tumors because of their localised nature and mass effect. However, the fact that they consistently lack chromosomal alterations speaks against its true neoplastic nature [3]. Hemangioma is one of the most common soft tissue tumors (7% of all benign tumors). Most hemangiomas are superficial lesions that have a predilection for the head and neck region, but they may also occur internally, notably in organs such as the liver [4]. Cavernous hemangioma is a term widely used to describe a solitary venous-lymphatic malformation. It is the most common vascular lesion of the orbit.1It is also the most common intraorbital tumor found in adults. Although histologically benign, they can encroach on intraorbital or adjacent structures and be considered anatomically or positionally malignant [5,6]. Most of these tumors are unilateral and can increase intraorbital volume with a resultant mass effect. Visual acuity or

field compromise, diplopia, and extraocular muscle or pupillary dysfunction can result from compression of intraorbital contents. The morbidity associatedwith orbital cavernous hemangioma is the threat of compressive optic neuropathy, extraocular muscle dysfunction, and cosmetic disfigurement [5,6]. The tumor most commonly affects middle aged females. 7 Most common presenting feature is a slowly progressive proptosis. Extraocular muscle impairment and impaired visual function are seen with large lesions and with lesions located at the orbital apex. Clinically, these tumors are soft and do not change in size with the Valsalva maneuver or with coughing, straining, or change in the head position [8].

Most frequent locations are retrobulbar muscle cone, especially the lateral aspect of the intraconal space. However, a small minority (<10%) of these lesions are extraconal [8]. Cavernous malformations usually are solitary and most often occur in the lateral aspect of theretrobulbar intraconal space. However, few multifocal lesions have also been reported. They are rarely intramuscular. They uncommonly involve the orbital apex, but when they do, they may cause monocular vision loss due to the compression of blood vessels that supply the optic nerve. They occasionally (5%–10%) extend intracranially through the superior orbital fissure. Bone remodeling is not uncommon, and intralesional calcification occurs occasionally. Associations with Maffucci syndrome and blue rubber bleb nevus syndrome have been reported [9].

However, very few extraconal lesions have been reported. Rama et al have reported a case of extraconal cavernous hemangioma with superomedial location. 2 Rizvi et al. have also reported multiple superomedial extraconal cavernous hemangiomas of the orbit associated with conjunctival cavernous hemangiomas [10].

Cavernous hemangioma typically appears as a well-circumscribed intraconal mass. Although most lesions are ovoid or round, larger lesions have lobulated margins. Larger lesions will distort surrounding structures, as opposed to lymphoma which molds around structures. CT shows homogeneous soft tissue density, and may show small calcifications or phleboliths. MR shows isointense T1 signal, bright T2 signal, dark internal septations, and a dark circumferential rim that represents a fibrous pseudo capsule [1].

Grossly, the specimen is nodular and soft. Microscopically, it iscomposed of large, dilated, blood-filled vessels lined by flattened endothelium. The vessels may be arranged in a roughly lobular arrangement or in a diffuse haphazard pattern.

The walls are occasionally thickened by an adventitial fibrosis, and inflammatory cells may be scattered throughout the stroma. Mature bone is occasionally present. Large, deep cavernous hemangiomas may undergo thrombosis, ulceration, and infection.

The thrombi may be seen in various stages of organization and recanalization, the latter including papillary endothelial hyperplasia (Masson lesion) [3,4].

Transnasal endoscopic resection of intraorbital tumors is feasible and may offer some advantages when compared to traditional approaches.

However, it is paramount to have specialized instruments such as long handpiece drills, goodcamera systems, and long bipolar forceps, as well as an experienced endoscopic surgeon, to control vascular lesions and potential life-threatening complications [5].

Conclusion

Cavernous hemangioma, though the most common vascular lesion in adults, is a rare tumor as such. Most frequent site for orbital cavernous hemangioma in intraconal. However, rarely, it can present as extraconal vascular lesion. Thus, possibility of cavernous hemangioma must be considered in such scenario.

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