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A CASE REPORT OF EXTRACONAL CAVERNOUS HEMANGIOMA OF ORBIT

Otorhinolaryngology

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ABSTRACT

Cavernous hemangioma is the most common benign noninfiltrative neoplasm of the orbit. Most cavernous hemangiomas are intraconal and lateral in location. They result from the new formation of vessels, proliferation of tissue components of the vessel wall, and hyperplasia of cellular elements ordinarily concerned with the genesis of vascular tissue.[1] Cavernous hemangiomas are well-circumscribed masses bounded by a fibrous pseudocapsule, without prominent arterial supply acounting for the relatively slow enhancement). They are composed of dilated large vascular spaces (thus cavernous) lined by flattened and attenuated endothelial cells 1,3. As flow is slow, and vascular spaces large, areas of thrombosis are common 3,4. Unlike the name 'hemangioma' suggests, these lesions may not be tumors as there is no cellular proliferation 3, but rather gradually enlarging vascular malformations and as such some authors prefer the term cavernous malformation.

KEYWORDS

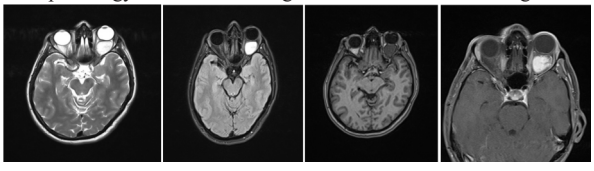
Extraconal, hemangioma, orbit

CASE REPORT

An 25-year-old woman presented with gradually progressive proptosis of the left eye with swelling of the superlateral quadrant of the left orbit. The swelling did not change in size with the Valsalva maneuver, coughing, straining, or change in head position. The cornea and sclera were normal.

USG revealed a well-encapsulated, compressible, echogenic mass lesion situated superolaterally in the extraconal space of the left orbit. On Doppler, the mass showed multiple vascular channels (both arterial and venous channels) suggestive of a vascular lesion . Small low-flow feeding arteries were seen. MRI showed an oval, encapsulated, and extraconal mass in the left orbit. The mass appeared isointense to orbital muscle on T1W and mildly hyperintense to orbital muscle on T2W images, with intense enhancement on post-gadolinium scans . There was medial displacement of the lateral rectus muscle anteriorly with inferomedial displacement of the globe. The intraocular muscles and optic nerve sheath complex showed normal signal intensity.

The tumor was excised using an anterior orbitotomy approach. Histopathology confirmed the diagnosis of a cavernous hemangioma.



Well defined round to ovoid T2/ STIR hyperintense mass in intraconal compartment of left eye with gradual and hamogenous post contrast enhancement Possibility of orbital haemangioma

DISCUSSION

Orbital cavernous hemangiomas are the most common benign masses in the adult orbit and the most common retrobulbar lesions in adults, after inflammatory pseudo-tumour and lymphoma. They are also considered as intraconal masses without optic nerve involvement (differential diagnosis of optic nerve sheath meningioma and optic nerve glioma). They consist in venous malformations of the orbit characterized by dilated, endothelial-lined vascular channels, with a variable amount of fibrous septations and a peripheral fibrous firm capsule. Because the blood flow within these vascular spaces is essentially stagnant, thrombosis is common, but spontaneous bleeding is rare. This is in distinction to cavernomas in the brain. They grow by thrombosis and subsequent recanalization of thrombosed sinusoids. They predominate in young to middle-aged women, but ranging from 10 to 60 years. Patients usually present with slowly progressive painless unilateral proptosis, but can also have diplopia, visual impairment and increased intra-ocular pressure with large lesions. They are almost always intraconal (80%), with extraconal malformations being uncommon. Is a well defined mass and may enlarge with a Valsalva maneuvre or during pregnancy. According to the grading system of vascular orbital lesions , cavernous hemangiomas are classified as type 3 - arterial flow lesions. They are considered low-flow AVM with small-caliber feeding arteries. However, these lesions are not easily identified at angiography. Support for an arterial supply is the fact that the majority of cavernous malformations arise in the lateral intraconal orbit in the area of the posterior ciliary artery.

The major role of MR in the evaluation of cavernous hemangiomas is to provide the precise anatomic delineation of the mass and its relationships to the optic nerve and other orbital structures. These lesions are surgically curable because they are encapsulated and do not recur if completely removed.

The differential diagnosis often includes a cranial nerve schwannoma because cranial nerves III, IV, V and VI have branches within the orbit. Both conditions are well-defined masses, hyperintense on T2-weighted images and enhance. It was shown on dynamic scanning that cavernous hemangiomas begin with a central spot or point of enhancement, which spreads peripherally (the contrast enters these lesions slowly, images obtained after injection usually show minimal or heterogeneous enhancement; it is more uniform on more delayed images), whereas orbital schwannomas usually enhance initially on the periphery. Both may remodel adjacent bone. If an intralesional (hyperintense on T2-weighted images) flow void is seen on T2-weighted images, another diagnosis should be considered, such as vascular hemangiopericytoma, venous varix, or an arteriovenous malformation.

Other masses that can occur in the orbit may be considered in differential diagnosis. Hemangiopericytoma is an uncommon vascular neoplasm with similar appearance (but with margins less well-defined) in neuroimaging studies, such as intense enhancement after contrast. Nevertheless, they are local aggressive and can destroy boneto extend beyond the orbit. Metastases can also occur, with a dense homogeneous stain in angiography. Orbital varix is also a vascular mass, uniformly enhancing, and distensible with Valsalva maneuvre. Treatment of the tumor is surgical excision. Complete excision is generally accomplished as the tumor is well encapsulated with relatively few feeding vessels.

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