GIANT CAVERNOMA OF THE ORBIT

Clinical and surgical considerations

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ABSTRACT - Cavernous hemangiomas of the orbit are benign, hamartomatous, vascular lesions, more frequent in middle-aged women, representing the most common benign primitive neoplasm of the orbit. Several therapeutic modalities and surgical approaches have been described, in order to preserve the normal orbital structures. We describe the case of a patient with a voluminous orbital cavernoma, approached by a fronto-orbito-zigotomy. A review on diagnostic modalities and surgical approaches is also presented.

KEY WORDS: cavernoma, surgical approach, fronto-orbito-zigotomy.

Cavernoma gigante da órbita: considerações clínico-cirúrgicas

RESUMO - Os hemangiomas cavernosos da órbita são lesões vasculares hamartomatosas benignas, mais frequentes em doentes do sexo feminino e de meia idade, constituindo a neoplasia benigna primitiva da órbita mais comum. Diversas modalidades terapêuticas e abordagens cirúrgicas têm sido descritas com a finalidade de preservar as estruturas orbitárias. Apresentamos o caso de uma doente com volumoso cavernoma da órbita esquerda, intervenção por abordagem fronto-órbito-zigomática. É efetuada ainda revisão das modalidades de diagnóstico radiológico e das diferentes estratégias cirúrgicas descritas na literatura.

PALAVRAS-CHAVE: cavernoma, abordagem cirúrgica, fronto-órbito-zigomática.

Cavernous hemangiomas of the orbit are benign vascular lesions accounting for approximately 5% of all primitive orbital neoplasms. They usually occur in adult-age, where they represent the most common vascular orbital tumor. They are more frequent in females and may grow during pregnancy. They differ from the brain cavernomas in their behavior and radiological features, and may coexist with venous angiomas in the same location suggesting a common origin and development. Multiple cavernomas of the orbit are rare entities that can recur after complete excision, and may exist with concurrent systemic vascular tumors. The posterior part of the orbit contains a number of important and delicate structures including the optic nerve, the ophthalmic artery and veins, the ocular muscles and their motor nerves, which makes the surgical access to this area quit difficult.

The case we present is illustrative.

CASE

39 years-old woman, presenting with recent, progressive, painless, proptosis in the absence of other clinical / neurological features.

MRI revealed a space occupying lesion in the left orbit, indicative of orbital cavernous hemangioma (Figs 1 and 2). The lesion was removed by a fronto-orbito-zigotomy extended to the medial wall of the contra lateral orbit. The procedure was uneventful and a cranioplasty with methylmetacrilate was performed for esthetical correction (Figs 3 to 6). The patient remained neurologically intact after the procedure.

Histopathological analysis revealed a cavernous angiomma of the orbit. Follow up MRI show no reminiscent, and, three years after surgery, the patient is still asymptomatic.

DISCUSSION

Histopathology – Vascular tumors of the orbit result from new formation of blood vessels, prolif-
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Fig 1. Coronal e axial T2 sequence of pre-operative MRI revealing expansive left orbital lesion, suggesting cavernoma.

Fig 2. Coronal e axial T1 sequence of post-operative MRI, showing resection of the lesion.

Fig 3. Fronto-orbito-zigotomy (zigotomy, pterional craniotomy and orbitotomy).

Fig 4. Left orbit after orbitotomy. The muscle elevator of superior eyelid, the frontal nerve (branch of the ophthalmic division of the trigeminal nerve) and the cavernoma can be seen.

Fig 5. Removal of the cavernoma after opening the periorbit and retracting the orbital structures.

Fig 6. After removal of the cavernoma, the muscles elevator of superior eyelid and superior rectus can be seen back in their anatomical position.

Operation of tissue components of the vessel wall and hyperplasia of cellular elements ordinarily concerned with genesis of vascular tissue. They are well delimitated benign vascular hamartomas, formed by sinusoidal vascular channels with irregular walls.

Clinical features – Painless and progressive proptosis associated or not with visual impairment, causing a variable degree of hyperopia, is the usual clinical presentation. Cranial nerves III - VI palsy and diplopia is uncommon.
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Diagnosis – Nearly all cases of orbital cavernoma can be correctly diagnosed by pre-operative neuroimaging study. The combined use of ultrasonography and computerized tomography (CT) may establish the nature, extent and location of tumors and vascular malformations of the orbit, and can be useful for selection of the surgical approach.

CT and magnetic resonance imaging (MRI) are of particular importance in the diagnosis of orbital vascular lesions. The improved image quality of CT and MRI along with dynamic CT angiography, MRI angiography, MRI venography, and multiphase dynamic contrast CT/MRI imaging, has proven useful to delineate and differentiate different orbital vascular lesions.

From the analysis of MRI appearance of an intraconal well-defined mass, with homogeneous signal, isointense to muscle on T1-weighted sequence, hyperintense on T2-weighted sequence, and progressive filling on gadolinium enhanced sequences, in a patient presenting with progressive, painless proptosis, the diagnosis of cavernous hemangioma is highly suggestive.

Scintigraphy with technetium Tc 99m (99mTc) labeled red blood cell, is a useful tool in the differential diagnosis of cavernous hemangioma from other orbital masses.

A complete neuroimaging study with computerized tomography, magnetic resonance and scintigraphy, enables the correct pre-operative diagnosis of vascular orbital lesions.

Treatment – In those cases presenting with visual impairment or significant exophthalmous, immediate treatment is indicated. The remaining, with minimal or non existing exophthalmous, can safely be followed by observation. There are cases in which the incomplete removal of cavernous hemangioma led to eventual involution of the reminiscent, with clinical relief.

Lateral orbitotomy has been widely employed for the removal of orbital tumors, being used in voluminous hemangiomas, especially those located in the lateral compartment of the orbit and lateral apex.

Anterior orbitotomy is useful in many cases, without significant complications and postero-inferior orbitotomy through the maxillary sinus can be used in small, well delimited lesions in the posterior and inferior orbit near the apex.

Resection of retro-bulbar tumors by transconjunctival approach is feasible, without relevant morbidity.

Endoscopic transethmoidal approach of the orbit is a minimally invasive surgery for retro-bulbar orbital neoplasm, leading to excellent cosmetic results with less bleeding. However, the access to intraconal lesions near the apex may be difficult.

Trans-frontal, trans-fronto-ethmoidal and trans-maxillary approaches have the disadvantage of possible injuries to a number of nontumor structures. Displacement of tumor lesions to the maxillary sinus has been used to prevent the traumatic and dangerous excessive traction of hemangiomas.

Lateral supraorbrow, trans-orbital sub-frontal and pterional approaches are employed for removal of tumors affecting the posterior two thirds of the orbit and tumors originating in or intruding into the optic canal.

Transcranial approaches offer an excellent surgical exposure and a good cosmetic effect and should be considered for big lesions located superiorly or medially to the optic nerve, especially those involving the apex.

Fronto-orbito-zigotomatic approach leads to excellent functional results, being of particular importance in deep lesions and in those with intracranial component. The possibility of anterior clinoidectomy and opening of the optic canal and superior orbital fissure, allows a wider surgical view and a safe transposition of the optic nerve.

Sclerosing endovascular therapy of low flow vascular lesions of the orbit has been advocated as an alternative to surgery in selected cases.

Cryosurgical extraction of cavernous hemangioma of the orbit, guided or not by stereotaxy has been described, but is not indicated for most orbital tumors.

The use in orbital surgery of neuronavigation systems offers three-dimensional coordinates of lesions, allowing a safer, controlled surgery.

Stereotactic radiosurgery is another treatment modality available for orbital and cavernous-sinus lesions.

In conclusion, cavernous hemangiomas of the orbit are benign lesions that usually occur in middle-aged women, presenting with progressive, painless proptosis. Due to improvement of neuroimaging techniques, it’s possible to obtain a correct pre-operative diagnosis in almost every case of vascular orbital lesions.

Despite of the employment of minimally invasive procedures in the treatment of these lesions, the authors consider that fronto-orbito-zigotomy is irre-
placeable in voluminous lesions of the two posterior thirds of the orbit, allowing an excellent cosmetic result with minimal morbidity.

REFERENCES


