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GIANT CAVERNOMA OF THE ORBIT

Clinical and surgical considerations

Feres Chaddad Neto[1], Armando Lopes[2], Mario Machado Filho[3], Aguinaldo Catanoce[3], José Maria Campos Filho[4], Evandro de Oliveira[5]

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 volumous 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, intervencionado por abordagem fronto-órbito-zigomática. É efetuada ainda revisão das modalidades de diagnóstico imageológico 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[1]. They usually occur in adult-age, where they represent the most common vascular orbital tumor[2]. They are more frequent in females and may grow during pregnancy. They differ from the brain cavernomas in their behavior and radiological features[3], and may coexist with venous angiomas in the same location suggesting a common origin and development[4]. Multiple cavernomas of the orbit are rare entities that can recur after complete excision, and may exist with concurrent systemic vascular tumors[5]. The posterior part of the orbit contains a number of important and delicate strutures 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 angioma 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-

[1]Neurocirurgião do Instituto de Ciências Neurológicas, São Paulo, Brasil (ICNE); Neurocirurgião-Assistente da Disciplina de Neurocirurgia do Departamento de Neurologia da Universidade de Campinas, Campinas SP, Brasil (UNICAMP); [2]Médico Residente do Centro de Neurocirurgia de Coimbra, Centro Hospitalar de Coimbra, Portugal (CHC); [3]Médico do Instituto de Ciências Neurológicas, São Paulo, Brasil (ICNE); [4]Médico Residente de Neurocirurgia da Faculdade de Ciências Médicas da Universidade Estadual de Campinas, Campinas SP, Brasil (UNICAMP); [5]Diretor do Instituto de Ciências Neurológicas, São Paulo, Brasil (ICNE); Professor e Chefe da Disciplina de Neurocirurgia da Faculdade de Ciências Médicas da Universidade Estadual de Campinas, Campinas SP, Brasil (UNICAMP).

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Dr. Feres Chaddad Neto - Praça Amadeu Amaral 27 / 5° andar - 01327-010 São Paulo SP - Brasil. E-mail: fereschaddad@hotmail.com

Fig 1. Coronal e axial T2 sequence of pre-operative MRI revealing expansive left orbital lesion, suggesting cavernoma.

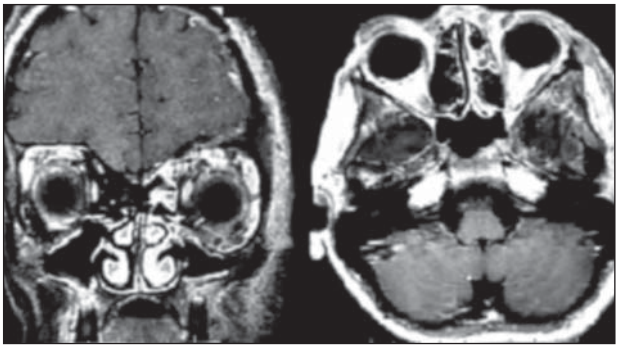

Fig 2. Coronal e axial T1 sequence of post-operative MRI, showing resection of the lesion.

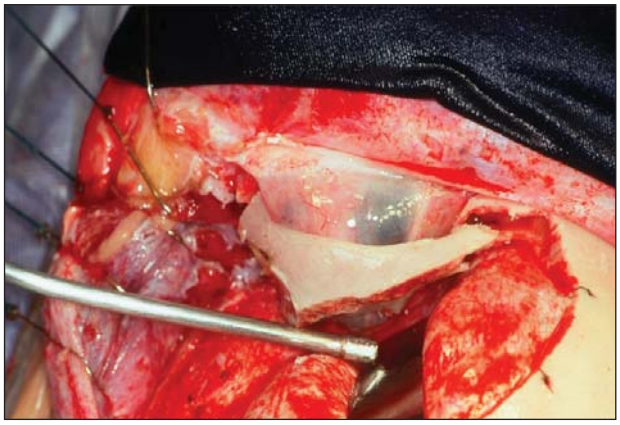

Fig 3. Fronto-orbito-zigotomy (zigotomy, pterional craniotomy and orbitotomy).

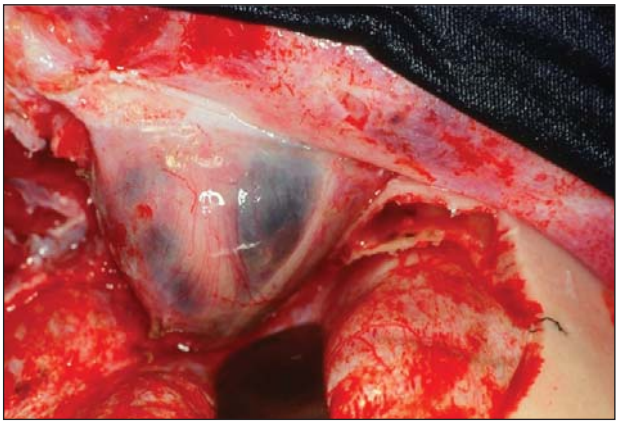

Fig 4. Left orbit after orbitotomy. The muscle elevator of superior eyelid, the frontal nerve (branch of the ophthalmic division of the trigeminal nerve) and the cavernoma can be seen.

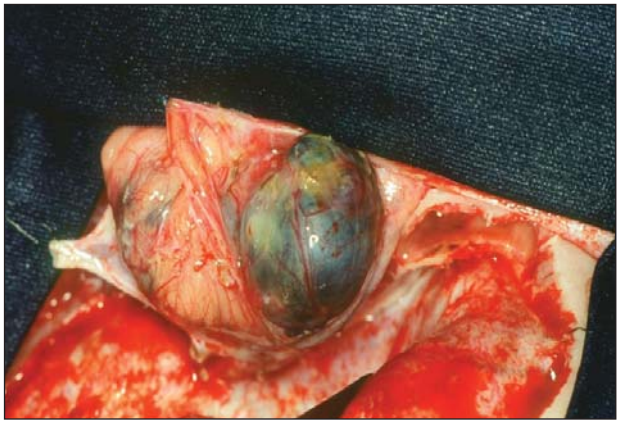

Fig 5. Removal of the cavernoma after opening the periorbit and retracting the orbital structures.

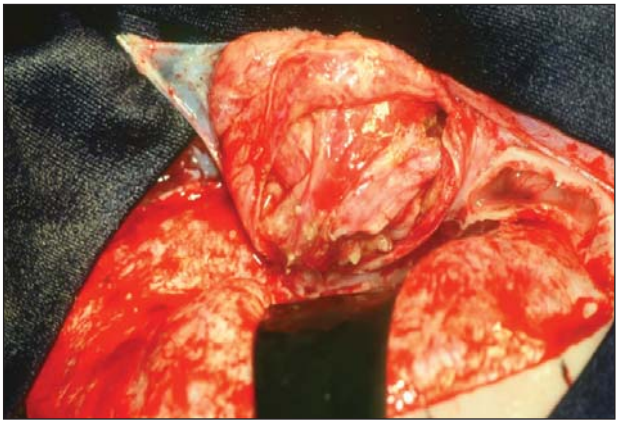

Fig 6. After removal of the cavernoma, the muscles elevator of superior eyelid and superior rectus can be seen back in their anatomical position.

eration of tissue components of the vessel wall and hyperplasia of cellular elements ordinarily concerned with genesis of vascular tissue[6]. 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[7]. Cranial nerves III - VI palsy and diplopia is uncommon.

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 approache[8].

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[9].

From the analysis of MRI appearance of a 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[10].

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[11, 12].

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[13]. The remaining, with minimal or non existing exophtalmous, 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[14].

Lateral orbitotomy has been widely employed for the removal of orbital tumors, being used in volumous hemangiomas, especially those located in the lateral compartment of the orbit and lateral apex[15].

Anterior orbitotomy is useful in many cases, without significant complications and postero-inferior orbitotomy through the maxillary sinus[16] 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[17].

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[18,19].

Trans-frontal, trans-fronto-ethmoidal and transmaxillary approaches have the disadvantage of possible injuries to a number of nontumor structures [19]. Displacement of tumor lesions to the maxillary sinus has been used to prevent the traumatic and dangerous excessive traction of hemangiomas[20].

Lateral suprabrow[21], 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[22].

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[13].

Fronto-orbito-zigotomatic approach leads to excellent functional results, being of particular importance in deep lesions and in those with intracranial component[23]. The possibility of anterior clinoide removal and opening of the optic channel 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 a alternative to surgery in selected cases[24].

Cryosurgical extraction of cavernous hemangioma of the orbit, guided or not by stereotaxy has been described, but is not indicated for most orbital tumors[25,26].

The use in orbital surgery of neuronavigation systems offers three-dimensional coordinates of lesions, allowing a safer, controlled surgery[27].

Stereotactic radiosurgery is another treatment modality available for orbital and cavernous-sinus lesions[28].

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 volumous lesions of the two posterior thirds of the orbit, allowing an excellent cosmetic result with minimal morbidity.

REFERENCES

1. Ohtsuka K, Hashimoto M, Suzuki Y. A review of 244 orbital tumors in Japanese patients during a 21-year period: origins and locations. Jpn J Ophthalmol 2005;49:49-55.
2. Gunalp I, Gunduz K. Vascular tumors of the orbit. Doc Ophthalmol 1995;89:337-345.
3. Acciarri N, Giulioni M, Padovani R, Gaist G, Pozzati E, Acciarri R. Orbital cavernous angiomas: surgical experience on a series of 13 cases. J Neurosurg Sci 1995;39:203-209.
4. Kodama T, Tane N, Ohira A, Matsuoka Y, Maruyama R. Concomitant cavernous hemangioma and venous angioma of the orbit. Jpn J Ophthalmol 2004;48:415-417.
5. Limawararut V, Davis G, Crompton J, Leibovitch I, Selva D. Recurrent multiple cavernous hemangiomas of the orbit in association with systemic tumors. Am J Ophthalmol 2006;141:943-945.
6. Hejazi N. Frameless image-guided neuronavigation in orbital surgery: practical applications. Neurosurg Rev 2006;29:118-122.
7. D'Hermies F, Elmaleh C, Mourier K, et al. Cavernous hemangioma of the orbit. Klin Monatsbl Augenheilkd 1992;201:291-301.
8. Xiao LH, Lu XZ, Wei H. Diagnosis by ultrasonography and CT scan in orbital vascular tumors and malformations. Zhonghua Yan Ke Za Zhi 2004;40:364-367.
9. Ansari SA, Mafee MF. Orbital cavernous hemangioma: role of imaging. Neuroimaging Clin N Am 2005;15:137-158.
10. Thorn-Kany M, Arrue P, Delisle MB, Lacroix F, Lagarrigue J, Manelfe C. Cavernous hemangiomas of the orbit: MR imaging. J Neuroradiol 1999;26:79-86.
11. Deol AK, Terry JE, Seibert DA, Solanki HP, Chertow BS. Hemangioma of the apical orbit diagnosed by radionuclide imaging. Optom Vis Sci 1994;71:57-59.
12. Polito E, Burroni L, Pichierri P, Loffredo A, Vattimo AG. Technetium tc 99m-labeled red blood cells in the preoperative diagnosis of cavernous hemangioma and other vascular orbital tumors. Arch Ophthalmol 2005;123:1678-1683.
13. Scheuerle AF, Steiner HH, Kolling G, Kunze S, Aschoff A. Treatment and long-term outcome of patients with orbital cavernomas. Am J Ophthalmol 2004;138:237-244.
14. Henderson JW, Farrow GM, Garrity JA. Clinical course of an incompletely removed cavernous hemangioma of the orbit. Ophthalmology 1990;97:625-628.
15. Arai H, Sato K, Katsuta T, Rhoton AL Jr. Lateral approach to intraorbital lesions: anatomic and surgical considerations. Neurosurgery 1996; 39:1157-1162.
16. Kennerdell JS, Maroon JC, Celin SE. The posterior inferior orbitotomy. Ophthal Plast Reconstr Surg 1998;14:277-280.
17. Hassler W, Schaller C, Farghaly F, Rohde V. Transconjunctival approach to a large cavernoma of the orbit. Neurosurgery 1994;34:859-861.
18. Tsirbas A, Kazim M, Close L. Endoscopic approach to orbital apex lesions. Ophthal Plast Reconstr Surg 2005;21:271-275.
19. Karaki M, Kobayashi R, Mori N. Removal of an orbital apex hemangioma using an endoscopic transethmoidal approach: technical note. Neurosurgery 2006;59:159-160.
20. Kosaka M, Mizoguchi T, Matsunaga K, Fu R, Nakao Y. Novel strategy for orbital tumor resection: surgical "displacement" into the maxillary cavity. J Craniofac Surg 2006;17:1251-1258.
21. Maus M, Goldman HW. Removal of orbital apex hemangioma using new transorbital craniotomy through suprabrow approach. Ophthal Plast Reconstr Surg 1999;15:166-170.
22. Marchal JC, Civit T. Neurosurgical concepts and approaches for orbital tumours. Adv Tech Stand Neurosurg 2006;31:73-117.
23. Akiyama H, Kondoh T, Suzuki H, Nakamura M, Ehara K, Tamaki N. Usefulness of fronto-orbito-zygomatic approach for intraorbital tumors: report of 31 cases. No Shinkei Geka 1997;25:913-917.
24. Schwarcz RM, Ben Simon GJ, Cook T, Goldberg RA. Sclerosing therapy as first line treatment for low flow vascular lesions of the orbit. Am J Ophthalmol 2006;141:333-339.
25. Gdal-On M, Gelfand YA. Surgical outcome of transconjunctival cryosurgical extraction of orbital cavernous hemangioma. Ophthalmic Surg Lasers 1998;29:969-973.
26. Papalkar D, Francis IC, Stoodley M, et al. Cavernous haemangioma in the orbital apex: stereotactic-guided transcranial cryoextraction. Clin Experiment Ophthalmol 2005;33:421-423.
27. Rosca TI, Pop MI, Curca M, et al. Vascular tumors in the orbit: capillary and cavernous hemangiomas. Ann Diagn Pathol 2006;10:13-19.
28. Thompson TP, Lunsford LD, Flickinger JC. Radiosurgery for hemangiomas of the cavernous sinus and orbit: technical case report. Neurosurgery 2000;47:778-783.