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CASE REPORT

VASCULAR ORBITAL TUMORS AT THE EXTREMES OF THE AGE SPECTRUM

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Abstract

Vascular orbital lesions are rare and, due to the controversy surrounding their origin, frequently difficult to diagnose. Studies showed that approximately 10% of orbital space-occupying lesions are of vascular origin. The most frequent are capillary hemangioma in children and cavernous hemangioma, which, although congenital, reveals itself in adults.
Two cases of vascular tumors in patients, at the extremes of the age spectrum are presented.
Keywords: orbit, hemangioma, infant

Introduction

Vascular tumors in the orbit are rare and due to controversies related to etiology, nomenclature and classification, they represent a diagnostic dilemma [**1**]. The most frequently encountered are capillary hemangioma, which constitutes approximately 24% of all vascular lesions and orbital cavernous hemangioma, which represents 26% [**2**]. Being the most common, the data in this article will refer only to these two entities.

Capillary hemangioma is the most common vascular tumor in children, with an incidence of 5.6% [**1**]. They are more common in women, occurring immediately after birth and usually regressing in 5-7 years. Most of them are extraconal and can be superficial or deep, the latter usually extending through the optical canal or the superior orbital fissure intracranial. Through its position, capillary hemangioma can lead to proptosis, globe displacement and amblyopia. Complications are rare and include bleeding, thrombosis, optic nerve compression, bone remodeling. Histologically, it has no capsule and is composed of lobules separated by fibrous septa [**3**]. For small lesions, the therapeutical approach is observation, intralesional corticosteroid injections, systemic interferon and systemic beta-blocker [**2**]. The administration of local corticosteroid injection is associated with severe side effects such as central retinal artery or vein obstruction, retinal embolism, suppression of adrenal glands or local hypopigmentation.

Systemic therapy with beta-blocker (propranolol) has been promoted in recent years with good and rapid results and with fewer complications. Treatment should be continued for 8-12 months, doses of 2-3mg/ kg divided in two administrations, under the supervision of a pediatrician. Side effects can be bradycardia, hypotension and hypoglycemia but they can be controlled if the patient is closely monitored during the treatment. There are several risk factors that predispose to the occurrence of these complications: premature patient, age below 3 months and asthma [**4**].

In the case of large lesions, with proptosis and long-term risk of amblyopia, surgical excision is recommended.

Cavernous hemangioma is the most common vascular tumor in adults. It is more common in women between 18 and 72 years [**5**] and presents as slowly growing masses that may lead in time to painless proptosis. Other signs at presentation are pain, eyelid edema, diplopia, palpable lesion and transient episodes of low visual acuity. They are solitary tumors that usually appear in the retro bulbar space. Rarely, they can extend to the intracranial level. Histologically, it has a fibrous pseudo capsule and is relatively well-defined [**2**]. Surgical excision is recommended in cases with proptosis, globe displacement with diplopia and optic nerve compression.

Clinical Cases

In the last 5 years we have encountered numerous cases of orbital hemangioma, both capillary and cavernous, all of which have been successfully excised. We present only two vascular orbital tumors in patients who were at different ends of the age spectrum, tumors that were large, extended deep in the orbit and required laborious surgery.

The first case is a 6-month-old girl who presented with a right orbital mass since she was 1 month old, mass that had grown rapidly in size. The clinical exam revealed a reddish subconjunctival lesion, highly adherent to the overlying tissue that then covered the temporal half of the pupil. The MRI exam showed an orbital mass of approximately 1,5 cm, situated superior and temporal, with a clear boundary between it and the globe and adjacent to the lateral rectus muscle (**Fig. 1**).

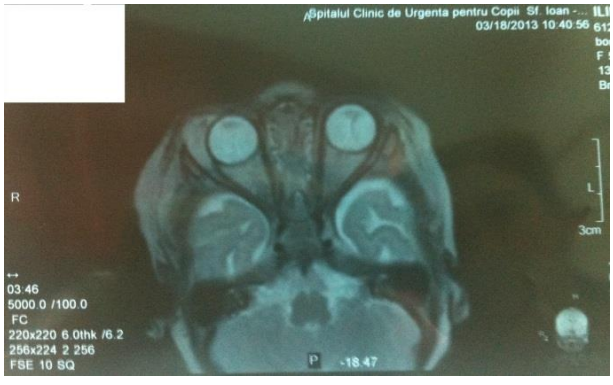

Fig. 1 MRI exam showing an orbital mass of approximately 1,5 cm

During surgery we found a 1.5 cm mass, highly friable and well vascularized, situated in contact with the lateral rectus (**Fig. 2**).

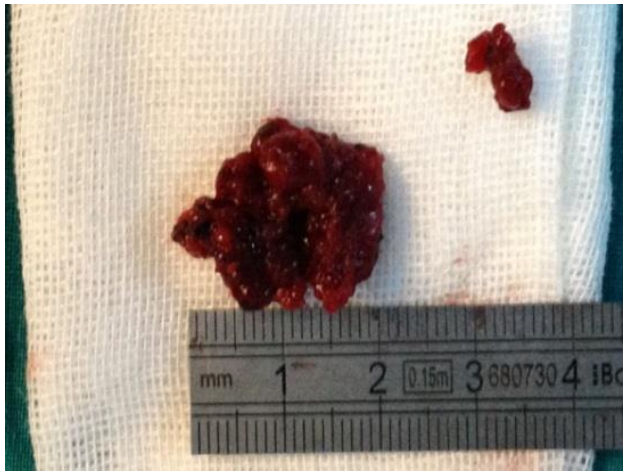

Fig. 2 1.5 cm mass, highly friable and well vascularized

After surgery, the patient presented minimal chemosis and good ocular motility (**Fig. 3**). The histopathological exam revealed a capillary hemangioma. The mother mentioned that her other daughter had a capillary hemangioma on the nose.

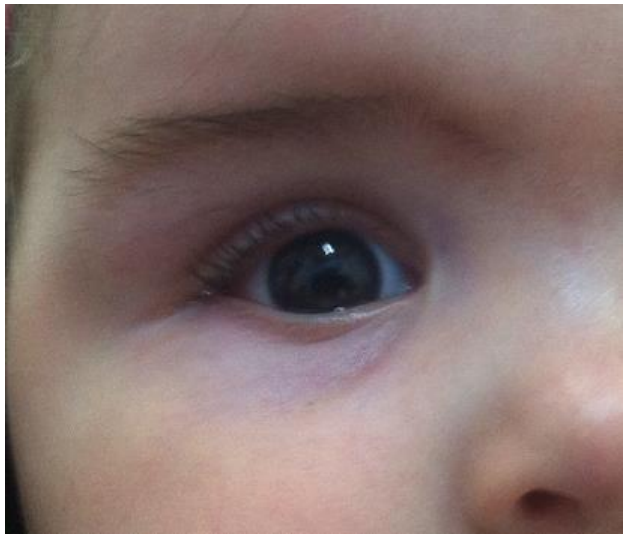

Fig. 3 Post-surgery image: minimal chemosis and good ocular motility

The second case was a 53-year-old male who presented with an orbital mass in the left infero-temporal quadrant, of approximately 25

years, that grew slowly and determined proptosis and globe deviation 7 years before. The patient also had transient diplopia in primary gaze and permanent diplopia in lateral gaze (the patient's compliance to treatment was very low, he had been scheduled for surgery twice in the previous years and did not show). The clinical exam showed proptosis, limited motility laterally, orbital mass in the left infero-temporal quadrant, purple and rather firm when palpated (**Fig. 4**).

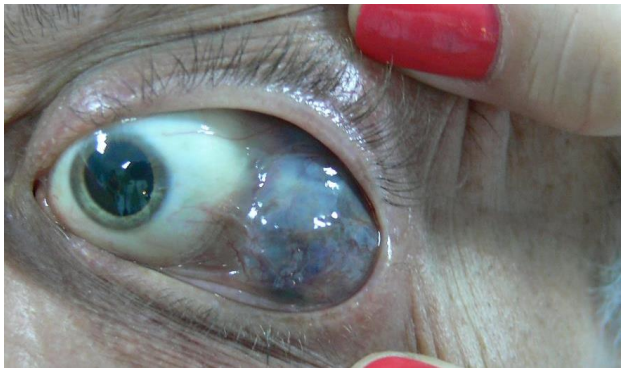

Fig. 4 Clinical exam: proptosis, limited motility laterally, orbital mass in the left infero-temporal quadrant, purple and rather firm when palpated

The MRI exam revealed a left multilobulated nodule, 32 mm in length, intra and extraconal, lateral and inferior, that infiltrated the anterior 2/ 3 of the lateral rectus muscle (**Fig. 5**).

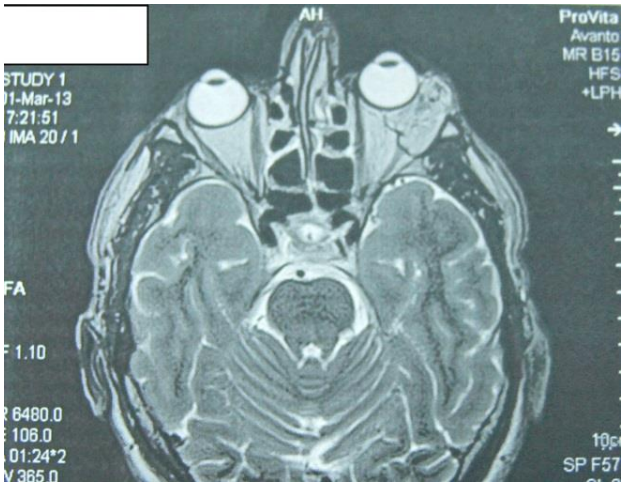

Fig. 5 MRI: left multilobulated nodule, 32 mm in length that infiltrated the anterior 2/ 3 of the lateral rectus muscle

During surgery, the conjunctiva was lifted in the infero-temporal fornix, a lateral canthotomy was performed to get a better exposure of the tumor and the mass was excised entirely, without damaging the lateral rectus. The histopathological exam revealed a cavernous hemangioma (**Fig. 6,7**).

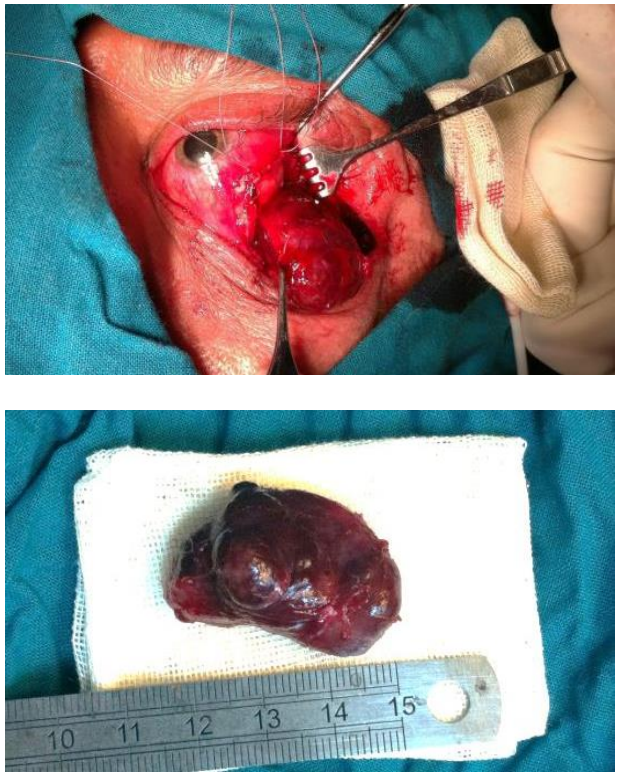

Fig. 6,7 Cavernous hemangioma

After surgery, the patient presented chemosis and eyelid edema, the same visual acuity as before surgery, the globe's lateral motility improved and currently he has no diplopia (**Fig. 8**).

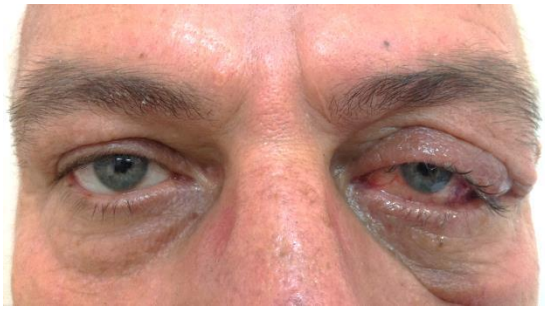

Fig. 8 Post-surgery state: no diplopia

Conclusions

Regarding hemangiomas in children, surgery is required when the mass is large enough to determine proptosis, globe compression or occlusion of the visual axis. In our case, due to the tumor's rapid growth, we decided to perform the surgery early in order to avoid these complications, mainly amblyopia by blocking the visual axis.

In the second case, the tumor's growth was slow (the size of the tumor was partly due to the patient's non-compliance) and the surgery was performed because of the diplopia and the esthetic appearance, actually the main reason the patient decided to present for surgery. Also, it should also be pointed out that, although the cavernous hemangioma is more frequent in women, our patient was male.

In both cases, and usually anytime we are dealing with an orbital mass, the imaging exams (MRI, CT) are essential in order to choose the right surgical approach and to diminish the complications, both during surgery and after.

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