Orbital varicose vein, exophthalmos or enophthalmos? A case report

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I. Introduction:

The term orbital tumours covers a wide spectrum of benign and malignant pathologies, affecting the orbital components or developed in contact with them. They are discovered by chance or sought during the workup of a general illness or in the presence of orbital signs (exophthalmos, pain, etc.). The classification based on anatomopathology is fairly widely accepted, and includes capillary haemangioma, cavernous haemangioma and haemolymphangioma. Although orbital varices are not tumours, it seems important to discuss this pathological entity because of the diagnostic problems it can pose and its similarity to haemolymphangiomas. Unilateral exophthalmos is most often due to an intra-orbital expansive process. Orbital varices, considered to be vascular tumours, are due to proliferation and dilation of intraorbital venous elements. They account for 2% of expansive orbital processes (1). We will discuss the difficulties of diagnosing orbital varices in our case report.

Clinical case:

This is a 65-year-old man with no particular pathological history who came to our consultation with a sensation of right exophthalmos that was bothersome and recurrent when prostrating during prayer, and had been evolving for more than 5 years. The ophthalmological examination revealed bilaterally corrected visual acuity of 9/10, and right-sided enophthalmos (Figure 1) which changed into painless exophthalmos when the head was tilted downwards (Figure 2). The rest of the eye examination was normal, with an IOP of 16 mmHg in the right eye and 15 mmHg in the left. An orbital MRI (Figure 3) was ordered, revealing dilatation of the right inferior ophthalmic vein and patent homolateral cavernous sinus veins. Additional scans, performed prone, showed further dilatation of the inferior ophthalmic vein with grade 2 exophthalmos. Given the absence of complications, a conservative approach was adopted with annual monitoring.

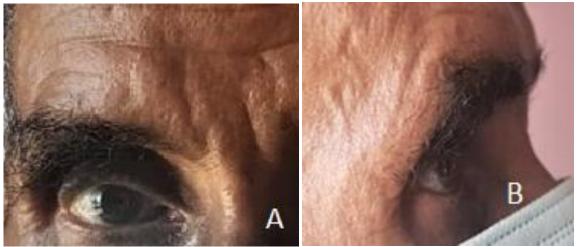


Fig. 1: Enophthalmos

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Fig. 2: Exophthalmos when the head is down

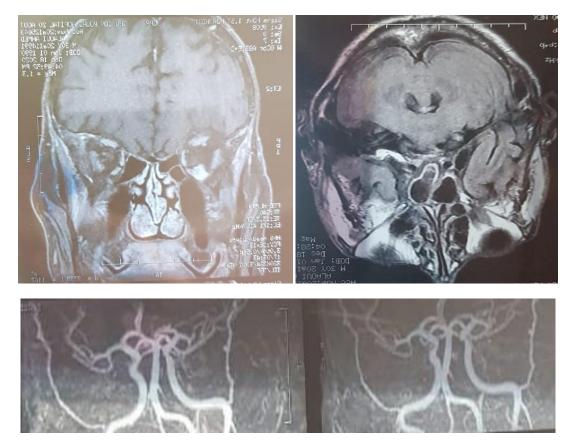


Fig. 3: Dilatation of the right inferior ophthalmic vein and patent homolateral cavernous sinus veins

II. Discussion:

Vascular anomalies account for a significant proportion of orbital tumours [2]. Orbital varices may be congenital, post-traumatic or secondary to an arteriovenous fistula. Most often unilateral, they often manifest as intermittent exophthalmos of a postural nature, aggravated by physical effort, manoeuvres inducing cephalic venous hyperpressure or the proclivity position, which was the case for our patient [3]. Attacks of exophthalmos are short and must be ruled out, in particular the dreaded carotid-cavernous fistula, by looking for a thrill. Only MRI angiography will confirm the diagnosis of orbital varices. Imaging is used to establish the diagnosis by comparing decubitus and procubitus views. It shows a formation that increases in volume with the valsalva manoeuvre or when the patient is in the prone position, and that is strongly enhanced with the presence of suggestive calcifications: phleboliths. Enophthalmos associated with orbital varices is rare but was found in our patient, probably explained by the expansion of orbital volume and atrophy of orbital fat [4]. Surgical treatment is indicated in the event of complications such as severe exophthalmos with corneal exposure, optic nerve

compression or intolerable pain [5], and consists of surgical removal and/or embolisation. The evolution of orbital varicose veins is either towards an increase in volume, thrombosis or even transformation into intravascular endothelial hyperplasia, or rarely towards spontaneous regression, which requires regular monitoring even in the absence of serious complications [6]. In our patient, given the absence of complications, we decided to institute annual monitoring.

III. Conclusion:

Always believe your patient. We should not rely on the slit lamp examination alone, which proved normal in our patient and which could lead to a delay in diagnosis; questioning is a crucial stage that should not be underestimated. Intermittent exophthalmos is certainly the major symptom of orbital varices, but enophthalmos should also prompt us to look for this diagnosis.

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