Intermittent exophthalmos studied with computerized tomography

Report of two cases

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Two cases of intermittent exophthalmos are reported. In both instances, cerebral angiography and orbital venography failed to outline the lesion, which was clearly demonstrated with the aid of computerized tomography. An orbital varix was seen to be the cause of proptosis in one surgically verified case, whereas in the other this same diagnosis was suspected on the basis of the clinicoradiological findings. The etiology, clinical manifestations, and management of orbital varix are briefly discussed.

KEY WORDS · intermittent exophthalmos · proptosis · orbital varix · vascular malformation · computerized tomography

In most instances, intermittent proptosis is caused by orbital vascular abnormalities.2–7,9,10,13–19,22–24,26,27,29 Among these, orbital varix consisting of a pathological enlargement of one or several venous channels within the orbit has been reported as the most frequent anomaly.2–4,6,7,9,10,14–19,22–24,27,29 Thus, the terms "orbital varix" and "intermittent proptosis" have become almost synonymous in the literature. Much less commonly, this rare and striking symptom is caused by hyperplasia of the lacrimal gland associated with dacryolithiasis21 or highly vascularized orbital tumors, such as hemangioma or lymphangioma, which are liable to undergo periodic congestion.6,10,12,29 Differentiation between these tumors and true vascular anomalies may be nearly impossible on clinical grounds alone; orbital venography1,2,4–6,9,10,12,14–19,22,23,25 and cerebral angiography3,6,9,10,13,14,15,17–19 are essential for an accurate diagnosis. However, these techniques may be unsuccessful in demonstrating orbital varices, even after inducing cranial venous engorgement by means of a Valsalva maneuver.3,6,9,10,18,19,27

We are reporting the cases of two patients suffering from intermittent exophthalmos, who were recently studied by us with the aid of computerized tomography (CT). To our knowledge, these are the first two reported cases of intermittent proptosis studied with this technique.

Case Reports

Case 1

This 32-year-old man had been suffering for 13 years from intermittent proptosis of the left eye caused by bending the head, compression of the neck, and Valsalva maneuvers. The initial onset of this symptom was related to a fragment of emery striking the eye, which could not be localized by the attending ophthalmologist. Apart from the painless intermittent exophthalmos related to position, the patient reported two episodes of severe painful proptosis of the left eye accompanied by marked congestion of the eyelids. The first of these two sudden spontaneous attacks occurred 10 years before admission, and lasted for a week. At that time, carotid angiography and orbital venography were performed, with negative results. A similar attack, lasting for 2 days, caused the present admission to our hospital. The patient was in generally good health and denied intracranial noise or any other symptomatology.

On the day of admission, a nonpulsatile, nonreducible, painful exophthalmos with conjunctival edema and swelling of the eyelids was present on the left side. There was no venous congestion of the forehead, and auscultation of the periorbital region was negative. Because of severe eyelid swelling, the
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**Case 1**

This 43-year-old man was admitted to our Service complaining of intermittent proptosis of the left eye. For 4 years before admission, this symptom occurred whenever he bent forward. Proptosis, which was absent when the patient was upright, could also be easily triggered by pressure over the left side of the neck. The patient never experienced local pain, cranial noise, diplopia, or visual deficit, and there was no history of trauma.

General physical examination was normal. When the patient was upright, a slight enophthalmos was appreciated on the left side. Bending forward elicited a direct, nonpulsatile, and painless proptosis of the left eye which immediately disappeared when the patient returned to the upright position. Proptosis could also be triggered by compressing the left jugular vein. Eye movements, visual acuity, visual fields, and optic fundi were normal. Bruit, thrill, and pulsation of the ocular globes were absent. Routine laboratory tests were normal. X-ray films of the skull and orbits were also normal.

The CT scan was normal under basal conditions but, following sustained bilateral jugular compression, a mass in the left orbital apex with the density of blood was demonstrated (Fig. 2). The appearance of the lesion did not change in the postcontrast scan. Left carotid angiography was normal. Orbital venography was initially normal but, after the exophthalmos had been elicited by means of jugular compression, a small amount of dye remaining in the apex of the left orbit was visualized (Fig. 3). Treatment was not advised and the patient was discharged.

**Case 2**

This 43-year-old man was admitted to our Service complaining of intermittent proptosis of the left eye. For 4 years before admission, this symptom occurred whenever he bent forward. Proptosis, which was absent when the patient was upright, could also be easily triggered by pressure over the left side of the neck. The patient never experienced local pain, cranial noise, diplopia, or visual deficit, and there was no history of trauma.

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**Fig. 1. Case 1.** A: Plain computerized tomography (CT) scan obtained when the patient was suffering from the second attack of severe proptosis. A retrobulbar mass of blood density causing marked exophthalmos may be seen on the left side. The transverse diameter of the orbit is enlarged. B: The postcontrast CT scan shows a minimal enhancement of the lesion. C: Follow-up CT scan performed after the proptosis had resolved spontaneously. There are no abnormalities except for the enlarged transverse orbital diameter.
Discussion

Intermittent proptosis should be differentiated from recurrent exophthalmos which may be due to a variety of conditions, such as periodic orbital edema, recurrent orbital hemorrhage, recurrent emphysema, and recurrent inflammatory conditions. When a patient presents with a transient or intermittent exophthalmos of the "now you see it-now you don't type," he most likely has an orbital varix. Orbital cavernous hemangioma may produce intermittent exophthalmos, but it usually occurs in an older age group and causes slowly progressive proptosis. The presence of a pulsating component in a patient with intermittent proptosis suggests the existence of an arteriovenous malformation (AVM), particularly if a bruit is present. It must be borne in mind, however, that a venous malformation, even if it is entirely intraorbital, may produce pulsating proptosis by transmission of the cerebral pulsation to the orbital contents through an enlarged sphenoidal fissure or a defect in the orbital roof in patients with neurofibromatosis.

Since an undetermined proportion of patients with intermittent exophthalmos follow a benign course, and the number of surgically verified cases is scant, a definite classification of the orbital vascular abnormalities is still lacking and the true incidence of orbital varix is not known. Different origins of orbital varix have been recognized. A definite cause was first reported by Walsh and Dandy in a patient in whom the enlarged intraorbital venous channels were seen to be secondary to an AVM of the middle cranial fossa. Lloyd, et al. pointed out that neither of the reported etiologies may account for all types of varices found in the orbit; they classified them as primary and secondary. According to these authors, primary intraorbital varices are in most instances true congenital venous malformations which are usually manifest soon after birth or in early childhood. Traumatic varices in which the onset of symptoms is related to ocular trauma and varices associated to orbital hemangioma are also considered primary, whereas enlargement of the intraorbital veins caused by shunt in cases of carotid cavernous fistula or AVM are considered secondary. Sometimes, orbital varices are associated with venous abnormalities in the scalp, forehead, and hard palate, and occasionally they form part of the Klippel-Trenaunay-Weber syndrome.

Intermittent proptosis related to orbital varices has been reported among all age groups, although there is a greater preponderance in the second and third decades of life. Males and females seem to be affected equally, and in the great majority of cases the left eye is involved, as occurred in our two patients. The clinical picture is typical: when the patient is upright no ocular or orbital abnormalities are observed, and in some cases even a slight enophthalmos is appreciated which has been attributed to the atrophy of the orbital fat and interstitial tissue resulting from the intermittently raised intraorbital pressure. This symptom was observed in our Case 2, despite the fact that attacks of severe proptosis had never occurred. Rapid protrusion of the eye, which is characteristically induced by bending for-

FIG. 2. Case 2. A: Plain computerized tomography (CT) scan in basal conditions shows no abnormalities within the orbits. B: Control unenhanced CT scan following compression of the left jugular vein in the neck. A retrobulbar mass of blood density producing moderate exophthalmos may be appreciated on the left side. The appearance of the lesion did not change in the postcontrast scan. C: The lesion as seen in the coronal plane CT images.
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ward, lowering the head, coughing, or compressing the jugular veins, quickly disappears as soon as the head is held erect or the induced venous congestion is relieved. Apart from the position-induced exophthalmos, sudden episodic attacks of severe proptosis, sometimes accompanied by pain and diplopia lasting from a few seconds to several days, may ensue without any apparent reason. Two of these episodes occurred in our Case 1 and led to surgical intervention. In some patients, proptosis gradually progresses over the years, the episodes becoming more and more frequent. Spontaneous improvement is rare and, although this lesion is not life-threatening unless associated with a cerebral AVM, it may cause optic atrophy in some patients. Ophthalmoplegia may also result from repeated severe attacks.

Despite the fairly typical clinical picture of orbital varix, the radiological studies are essential to achieve a definite diagnosis. In most cases of primary orbital varices, the standard roentgenographic views are negative. When the venous anomaly is found at birth, or has been present for a long time, enlargement of the orbital diameters may result. More or less typical x-ray features are the presence of phleboliths in the orbit and prominent vascular markings in the ipsilateral frontal bone. At carotid angiography, displacement of the ophthalmic artery may be seen, but the varix itself is rarely visualized during the venous phase. Not only the presence of the orbital mass but its true nature are most likely detected by orbital venography, which may show two types of venous abnormality: a local sacculated dilatation resembling a venous aneurysm or a whole system of abnormal venous channels throughout the orbit. However, venography may result only in partial filling or no filling at all of the varix, as occurred in our Cases 2 and 1, respectively; in both cases the CT scan proved to be highly sensitive in detecting the lesion. Although the nature of the orbital processes can only be tentatively presumed with the CT scan, the value of this technique for determining both the extent and location of the intraorbital masses is widely recognized. In our Case 1, the CT scan showed that the proptosis was due to a mass of blood density occupying the retrobulbar space, with the minimal enhancement seen following contrast injection suggesting a slow blood transit within the lesion. The plain and contrast-enhanced follow-up scans coincident with the spontaneous improvement were completely normal, and we believe that such behavior is rarely displayed by any lesion other than an orbital varix. In Case 2, the CT scan was also normal under basal conditions and only following vigorous jugular compression could a mass of blood density with irregular margins be demonstrated behind the left ocular bulb. This mass, which did not enhance following contrast injection, disappeared, as did the proptosis when jugular compression was relieved and the increased venous pressure diminished. Despite the fact that orbital venography was inconclusive in this patient and the CT scan did not demonstrate the nature of the lesion, we believe that the CT image, added to the typical clinical picture, makes the diagnosis of orbital varix highly probable.

Treatment of patients with orbital varix remains controversial. Leaving aside cosmetic considerations, a surgical approach is recommended when recurring attacks of proptosis threaten visual function. Retrobulbar injections of sclerosing agents or ligature of the ophthalmic...
vein\textsuperscript{6,9,10,23,27} in the orbit have been employed in the past. However, these procedures are liable to damage the ocular globe.\textsuperscript{3,4,6,9,27} The best form of treatment is radical excision.\textsuperscript{3,6,9,10,22,26} Removal may be facilitated by electrically induced thrombosis.\textsuperscript{9} Sometimes total excision is difficult because the venous channels partially collapse and remain undetected.\textsuperscript{10,24} This probably occurred in our Case 1, but even incomplete removal may result in lasting subjective and objective improvement.\textsuperscript{10}

References


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