Large Varix of the Superior Ophthalmic Vein: Demonstration by Angular Phlebography and Removal by Electrically Induced Thrombosis

Case Report

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Intermittent exophthalmos is very rare. Birch-Hirschfeld saw only one case among 150,000 patients with ophthalmic disease. This condition is usually caused by a varix in the orbit. Since the first report by Schmidt, orbital varices have attracted considerable attention because of their prominent symptom of a transient or intermittent exophthalmos. A variety of therapies have been tried. Since Krayenbühl demonstrated an orbital varix in two cases by angular phlebography, this condition has received more attention from neurosurgeons.

The condition is characterized by pronounced and rapid protrusion of one eye when venous stasis is induced by bending the head forward, lowering, or forcibly turning it, hyperextension of the neck, coughing, forced expiration, or pressure on the jugular veins. The ocular protrusion disappears immediately when the head is erect and when induced congestion is relieved. Usually there is enophthalmos when venous congestion is relieved. It is always unilateral and usually left-sided. There has been considerable speculation on the etiology of this condition. Walsh and Dandy's reviews of the literature on intermittent exophthalmos suggested that large congenital varicosities in the orbit associated with an obstruction of the outflow of blood from the orbit may be a possible cause. However, they felt that an obstruction within the veins could not explain intermittent exophthalmos, because proptosis appeared promptly with jugular compression, showing that obstruction was not present in the venous channels.

Most cases become evident in mid-life or later so that, while a congenital structural predisposition undoubtedly exists, the varices may not develop until after maturity is reached.

The purpose of this paper is to report a case of an orbital varix which was diagnosed clinically, demonstrated by angular phlebography, and successfully treated by surgery through the transcranial approach with the aid of electrically induced thrombosis.

Case Report

The patient, a 60-year-old man, was admitted to Kyoto University Hospital on September 13, 1965, with the chief complaint of double vision and intermittent right-sided exophthalmos.

Four years earlier he had first noted exophthalmos when he flexed his head anteriorly with a rapid return to normal when he raised his head. The exophthalmos was associated with transient diplopia, but he never experienced any murmurs or pain in the eye. There was nothing remarkable in his past or family history.

Examination. The general physical examination was normal. The blood pressure was 132/80, and the pulse rate 60 per minute and regular. The patient had no neurological deficit except bilateral mild impairment of hearing. There was no disturbance of eye movements, but a moderate degree of exophthalmos appeared on the right side when he lowered his head for a few minutes in such a way that venous congestion of the head was induced.

Routine laboratory examinations of the blood, urine, blood chemistry, and liver function were normal. There was no abnormality in the electrocardiogram or x-ray examination of the chest and skull. Right carotid angiography revealed no abnormality. Orbital phlebography was performed on Oc-
Electrical Thrombosis for Varix of Ophthalmic Vein

October 6, 1965, through a catheter inserted into the right vena angularis; an intraorbital varix was well demonstrated (Fig. 1).

Operation. On Oct. 20, 1965, a right frontal craniotomy was performed through a coronal incision. The right orbital roof was exposed extradurally and removed by the usual unroofing technique. Orbital fatty tissue was carefully removed through the divided opening of the levator palpebrae superioris muscle, and the major portion of the hemangioma was exposed by using a dissecting microscope (Fig. 2). Because of difficulty in distinguishing the main trunk of the varix, treatment by electrically induced thrombosis was decided upon. A cathode needle was connected to the frontal muscle, and positive electrodes were inserted manually into the hemangioma. An electric current was run with an intensity of 5 mA for 10 to 20 minutes four times successively from different angles to induce thrombus formation. The size of the hemangioma was thus greatly reduced so that the next step was greatly facilitated. The vessel draining the hemangioma was identified and ligated, and the entire varix was removed without difficulty.

Postoperative Course. The postoperative course was uneventful except that the patient had persisting right ptosis and diplopia. Because muscle injury was suspected, an exploration was done 8 days after operation. There was edema of the orbital structures, but no injury to the muscles or nerves was found. The ptosis and diplopia gradually disappeared. An angiogram taken 35 days after operation showed complete disappearance of the varix. The patient was perfectly well and engaged in his regular occupation when last seen in March, 1967.

Discussion

Successful differentiation between intraorbital tumors, vascular malformations, or inflammatory changes is difficult by neuroophthalmic examination alone, even when aided by routine radiographic examinations. Demonstration of the ophthalmic artery is helpful in the diagnosis of lesions involving the arterial system, such as intraorbital aneu-
rysm. However, intraorbital masses that most frequently cause intermittent unilateral exophthalmos are venous tumors. In such cases arteriography cannot provide adequate information.

In orbital phlebography, the vessels are often masked by bony structures in the paraorbital areas, and the picture obtained is difficult to interpret. Phlebograms produced by the method of subtraction are helpful in the demonstration of the vessels.

In a majority of cases, intermittent exophthalmos is progressive. Even after a few attacks at long intervals optic atrophy and complete blindness of the eye may result. If the recurrences last for many years, blindness is not at all uncommon and has in fact occurred in about 15% of the cases reported. Although danger to life does not arise except when there are also cerebral varices, the prognosis should always be guarded owing to the danger of eventual optic atrophy. If, despite prophylaxis to avoid venous congestion, crisis does recur, more active measures may be necessary. Injection of sclerosing agents into the orbit, puncture and aspiration of the retrobulbar space, ligation of the superior ophthalmic vein in the orbit through a supraorbital incision, have been reported as successful means of treating this condition, but these procedures are technically not at all easy and may produce ophthalmoplegia. The best procedure, we consider, is phlebographic evaluation followed by transcranial exploration of the lesion and its total extirpation.

During the past 6 years, we have studied artificially induced thrombosis, especially that created by an electric current. It has been demonstrated that the imposition of a charge on the vessel wall to reverse normal polarity causes the creation of a thrombus near the positive electrode, and bleeding is thus controlled. On the basis of these experimental studies, a new method was designed to obliterate the lumen of an intracranial aneurysm by electrically induced thrombosis with a fine platinum “acupuncture” needle (0.2 mm in diameter). Our clinical experience indicated that an anodal current worked better in shrinking arteriovenous malformations before complete extirpation and in controlling bleeding by electrical hemostasis than in causing complete obliteration of the lumen. In the case reported here, shrinkage of the varix was also observed and total extirpation was accomplished without much difficulty or gross bleeding.

Summary

A large varix of the right superior ophthalmic vein associated with intermittent exophthalmos has been described. The varix was demonstrated by angular phlebography and verified at the time of operation. Removal was facilitated by electrically induced thrombosis.

References


