Intermittent exophthalmos

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The syndrome of intermittent exophthalmos classically consists of enophthalmos with the head in the upright position, and exophthalmos which develops rapidly in one eye (usually the left) when venous stasis is induced by bending or lowering the head, turning the head forcibly, hyperextension of the neck, coughing, forced expiration with or without compression of the nostrils, or pressure on the jugular veins. The condition is often referred to as orbital varix, varicocele, venous aneurysms, or arterio-venous aneurysm (Walsh and Dandy, 1944; Brauston and Norton, 1963).

Cases of intermittent exophthalmos have been reported by Birch-Hirschfeld (1930), Wissmann and Schulz (1922), Rumjanzewa (1930), Bartók (1931), de Petri (1935), Marchesini (1935), Hippert (1936), Muirhead (1936), Kraupa and Mendl (1936), Lipowitsch (1936), Petrov (1939), Spektor (1939), Giquéaux (1942), Poole (1942), Dunphy (1942), Roncs (1942), and Walsh and Dandy (1944).

Orbital varices constitute about 90 per cent. of cases of intermittent proptosis (Duke-Elder, 1952). Recurrent orbital haemorrhages may also cause intermittent proptosis (Meyer, 1898; Teillais, 1898; Gayet, 1899), and highly vascular orbital neoplasms, such as angioma (von Graefe, 1866; Hegner, 1915; Augstein, 1917; von Hippel, 1918), lymphangioma (Wintersteiner, 1898; Franklin and Cordes, 1924; Wolff, 1932), lymphoma-sarcoma (Perzewa, 1922), and retrobulbar venous cavernous haemangioma (Zeidler and Zetterström, 1959) are other lesions which may give rise to this condition.

Intermittent proptosis accompanied by chemosis may be caused by venous congestion; this is sometimes seen during menstruation (Krauss, 1910; Morelli, 1922; Kragh and Holm, 1923), during the strain of childbirth (Rumjanzewa, 1930), or during a paroxysm of crying in infants which leads to spasm of the orbicularis, and may even result in dislocation of the globe (Duke-Elder, 1952). The condition may also occur if thrombosis of the cavernous sinus reduces the efficiency of the venous return from the orbit.

Other rare causes of intermittent proptosis are periodic orbital oedema (particularly of the angioneurotic type), recurrent emphysema (Duke-Elder, 1952), intermittent ethmoiditis (Selkin, 1925; Benoit, 1926; Juler, 1932), and intermittent otitis (Eagleton, 1935).

Case report

A young woman aged 21 years came to the Out-patients Clinic of the Ophthalmic Hospital at Tanta, complaining of protrusion of the left eye on bending the head downwards or stooping, accompanied by pain in the eye and headaches. The condition had started gradually 2 years before, with protrusion of the left eye whenever the patient lowered her head, but recovery immediately the head was raised. Pain and headaches had begun to appear only 6 months previously and occurred whenever the eye protruded.

There was no history of trauma, cough, sinusitis, or fever, but the condition had first appeared after the patient's second labour.

Examination

The eyes were myopic, with a temporal myopic crescent in the fundus. The visual acuity was 1/60 in both eyes without glasses, and 6/24 with correction—10 D sph. in the right eye and —9 D sph. in the left.

The left eye showed enophthalmos when the head was upright (Fig. 1). Bending the head slightly forwards caused a moderate protrusion of the left eye (Fig. 2), and lowering the head downwards was immediately followed by a marked protrusion of the left eye (Table), which recovered when the head was raised. Lateral rotation of the head also produced protrusion of the left eye (Fig. 3). The proptosis was non-pulsatile and non-reducible. The movements of the eyeball were normal. Narrowing of the palpebral fissure was noticed during the episodes of protrusion (Fig. 4). There was no change in the fundus appearance during the protrusion. No dilated veins were evident on the lids or face.

FIG. 1 Enophthalmos in left eye when head is upright

FIG. 2 Moderate degree of protrusion of left eye, when head is bent slightly forwards

FIG. 3 Protrusion of left eye with lateral rotation of head

FIG. 4 Narrowing of left palpebral fissure during protrusion of left eye

Table Exophthalmometric measurements with the head in different positions

Position of head	Exophthalmometric measurement (mm. Hertel)	
	Right eye	Left eye
Upright	17	15
Bending slightly forwards	17	21
Lowering head downwards	18	25
Turning laterally	17	22

Investigations

Systemic and ear, nose and throat examinations revealed no abnormality. There was no evidence of any endocrine disturbance.

X rays of the skull, orbits, and paranasal sinuses appeared normal. The basal metabolic rate and blood picture were also normal. Arteriography and phlebograms of the orbital veins were not performed as facilities were not available.

Discussion

Krauss (1910) assumed that the presence of large congenital orbital varicosities and any obstruction to the drainage of orbital blood, either anteriorly or posteriorly, could produce intermittent exophthalmos.

Brauston and Norton (1963) stated that the exophthalmos was due to a "vascular lake" behind the globe, probably congenital or developmental in origin. This could lie entirely within the orbit, or be connected with the cranial cavity; it emptied in the upright position and filled with any obstruction in the venous drainage.

The condition is usually progressive, and the most frequent complaint, which is common in the second or third decade, is fullness, bulging, and pain in the involved eye together with dizziness and vertigo when the patient bends forwards, or when there is impairment of the venous return from the head (Walsh and Dandy, 1944; Brauston and Norton, 1963). Birch-Hirschfeld (1930) suggested that the presence of intracranial varices might account for these symptoms.

All the reported cases have been unilateral. Poole (1942) remarked that the left eye was involved ten times more than the right, and Reese (1935) pointed out that the left jugular foramen is frequently smaller than the right.

Birch-Hirschfeld (1930) observed the occurrence of intermittent exophthalmos in relation to the various positions of the head. A latent period of 5 seconds was followed by a protrusion of the eye when the head was bent forwards, which reached its maximum in 30 seconds. Lateral rotation of the head was also found to influence the position of the eye, owing to a degree of jugular constriction. Proptosis can also be produced by pressure over the jugular vein on the affected side (Walsh and Dandy, 1944). Mulder (1900) reported a case which was evident only when the patient wore a tight collar.

Enophthalmos is usually apparent only when the head is in the erect position, and is due to the absorption of the orbital fat from long-standing pressure of the vascular bed in the orbit (Walsh and Dandy, 1944).

Narrowing of the palpebral fissure during the episodes of exophthalmos was observed by Poole (1942), Rumjanzewa (1930), and Walsh and Dandy (1944). Pupillary dilatation has also been described by several observers, although Rumjanzewa (1930) observed narrowing of the pupil in her case.

Walsh and Dandy (1944) noticed overfilling of the retinal veins, which pulsed during the protrusion; but otherwise the fundus was usually normal. In no instance have there been dilated veins over the lids, face, and scalp, such as are commonly observed in cases of carotid-cavernous fistula and cirroid aneurysm. Pulsations of the eyeball are rare.

The involved eye in most cases remains normal, although optic atrophy was noticed by Birch-Hirschfeld (1930) in 14 per cent. of cases; this he claimed to be due to retrobulbar haemorrhage which was an occasional complication. Walsh and Dandy (1944) suggested that the optic atrophy could be due to long sustained-direct pressure of the vascular mass on the optic nerve.

X rays have usually been reported as negative. An enlarged superior orbital fissure was noted by Walsh and Dandy (1944), and an enlarged orbit by Jones (1961). The presence of phleboliths was reported by Kraupa and Mendl (1936), Teitgen and Benedict (1951), Hu Yung-Lin and Shih Chen-Jung (1954), and Hager (1958).

Arteriography, which has rarely been performed proved negative on two occasions (Uberall, Schweitzer, and Gonzalez, 1955; Zeidler and Zetterström, 1959). One case revealed an intraorbital arteriovenous aneurysm without intracranial extension (Jacas, Ley, and Campillo, 1959).

Phlebograms of the intraorbital veins have recently been carried out *via* the angular vein in a case of intermittent exophthalmos by Krayenbühl (1962), who noticed a "cherry-like enlargement of the superior ophthalmic vein".

The first surgical procedure was performed by Schimanowsky (1907), who clamped the superior ophthalmic vein in the back of the orbit, through an incision beneath the eyebrow. Other attempts to ligate the dilated veins in the orbit were performed by Löwenstein (1911), Germain and Weill (1927), Rumjanzewa (1930), and Brauston and Norton (1963). Serious complications were encountered in some cases, such as spontaneous severe haemorrhage (Löwenstein, 1911) and post-operative external ophthalmoplegia with enophthalmos (Germain and Weill, 1927).

Brauston and Norton (1963) advised that, if the orbital fissure was normal and arteriography and phlebography did not demonstrate any intracranial extension, the varix might be isolated and excised by an orbital approach, but if the sphenoidal fissure was enlarged or there was arteriographic evidence of intracranial extension, the transcranial approach of Walsh and Dandy (1944) should be preferred. They observed, however, that "it may be impossible to remove such a vascular anomaly without causing irreparable damage to the function of the globe".

The injection of sclerosing solutions into the orbital veins has been advocated by several authors (Raverdino, 1925; Hippert, 1936; Tanzi, 1948; Sená, 1955), but there is always the risk of serious sequelae if intracranial extensions exist (Brauston and Norton, 1963).

Summary

A case is reported of intermittent exophthalmos due to an orbital varicocele, which developed after labour.

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