Case Reports

The Pathological Basis for Postural (Intermittent) Exophthalmos*

Case Report

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Postural or intermittent exophthalmos is an uncommon condition with distinctive clinical characteristics. Although Walsh and Dandy and Brauston and Norton reviewed 133 cases, the pathological anatomy of postural exophthalmos has been identified and corrected in only one instance. This was a case reported by Walker. We are reporting an additional case in which craniotomy established the pathological basis and cured the condition. The patient had an unrelated abnormality not present in any previously reported case of postural exophthalmos, namely, a defective development of the orbit associated with multiple neurofibromatosis.

Terminology

Exophthalmos from most causes becomes stationary after full development. Two varieties of exophthalmos are not fixed, and their variability is expressed in the terms recurrent and postural (intermittent) exophthalmos.

Recurrence exophthalmos is characterized by the appearance and subsequent disappearance of exophthalmos on one or more occasions. Recurrent exophthalmos has been described in association with a variety of conditions such as ocular inflammation, rupture of orbital cysts, thrombosis of venous varices, hemorrhage, hemangiomas, and lymphangiomas.

Postural exophthalmos, also known as intermittent exophthalmos, is characterized by temporary protrusion of an eyeball during maneuvers which raise intracranial venous pressure above that present normally in the erect position. At rest with the head elevated, the affected eye assumes a normal position, or is enophthalmic. Lowering of the head, straining, movements of the head and neck causing obstruction of the jugular veins, and digital occlusion of the jugular veins produce exophthalmos within a matter of seconds. The enophthalmic eyeball recedes just as rapidly when the intracranial venous pressure returns to normal.

Case Report

T.H., a 34-year-old white man, was admitted to the Christ Hospital on October 3, 1964, because of intermittent protrusion of the left eye, accompanied by severe throbbing pain.

History. Cutaneous neurofibromas had been present since childhood. In 1961 he underwent surgery for removal of bilateral retroperitoneal pheochromocytomas discovered during the course of investigation for paroxysmal hypertension. Bilateral carotid arteriograms, performed because of generalized headaches, were normal at that time. The following year he first noticed protrusion of the left eyeball when bending forward, coughing, or straining. On these occasions, he was aware of pulsations which were sufficient to cause blurred vision. Soon after its appearance, exophthalmos became associated with throbbing pain.

The patient entered the University of Kentucky Hospital, Lexington, in November, 1963, because of headaches, pain in the left eye, and loss of hearing in the left ear. Areas of brown pigmentation and multiple neurofibromas were present over the body. The left eye, slightly enophthalmic in the erect position, protruded immediately and to a striking degree when the patient coughed, performed the Valsalva maneuver, or bent forward (Fig. 1). A mild perceptive deafness was detected in the left ear. An ovoid defect in the outer table of the skull,
The left eye is enophthalmic with the patient at rest in the erect position (left) and exophthalmic during performance of the Valsalva maneuver (right).

2.5×2 cm, was palpated directly behind and above the left mastoid process; it was not tender and did not pulsate.

A lumbar puncture, a brain scan with Hg 197, and a pneumoencephalogram were normal. Plain radiographs of the skull and orbits revealed defects of the greater and lesser sphenoid wings and orbital plate, absence of the left anterior clinoid process, thinning and slight outward bulging of the left temporal squama (Fig. 1), and a defect involving the outer table and diploe near the lower end of the left lambdoid suture.

Craniotomy was undertaken on December 5, 1963, for the purpose of covering the orbital defect with tantalum mesh or an acrylic plate. A mass of dilated veins was encountered in the region of the Sylvian fissure. Some of these vessels entered a greatly enlarged sphenoparietal sinus. No attempt was made to deal with the vascular malformation, and the procedure was terminated on the basis that symptoms did not justify the risk of proceeding further. The postoperative course was uneventful. In the following year, pain in the left eye became increasingly severe, and the patient sought treatment at Christ Hospital.

Examination. At the time of admission to Christ Hospital, the left eye pulsated faintly and was slightly enophthalmic in the erect position. Performance of the Valsalva maneuver produced immediate forward protrusion of the pulsating globe for a distance of 18–20 mm; the globe receded immediately when the maneuver was terminated. Digital compression of the right jugular vein caused prompt protrusion of the left globe. Compression of the left jugular vein had no effect. When the patient bent forward from the waist, the eye gradually protruded until the head reached the level of the heart at which point the protrusion increased abruptly. Each time exophthalmos was induced, the patient suffered sharp, throbbing pain in the involved eye, and vision in this eye became blurred.

Radiographs of the skull revealed a left frontotemporal craniotomy defect and the