A case of intermittent exophthalmos

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Intermittent exophthalmos was first described by Schmidt (1805) as a clinical entity in an infant and has since been recognized as a rare but striking condition, its rarity being evidenced from the paucity of reported cases. Two important papers relating to this topic have been the subject of a comprehensive review of the world literature in the first half of this century—the first by Birch-Hirschfeld in 1930 and the second by Walsh and Dandy in 1944, and since then scarcely anything more has been added to our knowledge of this condition that is not contained in these reports.

Brauston and Norton in 1963, adding two cases of their own to the number previously reported, brought the total up to 137. In recent years, a survey of the *Index Medicus* reveals a few more contributions to the list to which the authors wish to add this personal case.

CASE REPORT

The patient, G. B., was a 41-year-old woman whose chief complaint was a distressing feeling of fullness in the right eye in certain bodily positions which was immediately relieved on sitting or standing upright. She stated that at the age of 6 years, there was a sudden slight protrusion of the right eyeball which within two hours was normal again. At age 20, she was operated upon for a hemangioma over the right upper eyelid which first made its appearance at the age of 14 and had slowly increased in size since then. At 24, she noticed that the right eye would 'pop' out of its socket with any form of excitement, forward bending of the head, or when she walked into a warm room from the outside cold in the winter months, but, as she attained the erect posture, the eye swelling would disappear almost as rapidly as it proptosed and she would be comfortable again. This situation became slowly and progressively worse over the years, until at the age of 39 she noticed that lying in bed, and especially on her right side, produced a proptosis which became almost unbearable. Other than the feeling of fullness, there was no pain in the eye, no disturbance of vision, and no pulsations or unpleasant noises heard. The left eye was normal at all times. There was no history of trauma.

PHYSICAL EXAMINATION The patient was of normal build and average intelligence. Blood pressure was 120/70.

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General physical examination was within normal limits and, except for the following findings in the right eye, the neurological examination was unremarkable. In the erect posture of the head, inspection showed a slight enlargement of the right palpebral fissure ($R = 12$ mm; $L = 11$ mm). Enophthalmos was clearly evident ($R = 15.5$ mm; $L = 17$ mm). There was a small varicosity near the inner canthus of the eye. The pupils reacted normally to light and accommodation. Eye movements were normal. There was no diplopia, nystagmus, or ptosis. When light was thrown on the right eye, a faint pulsation was visible in the eye and upper lid, but it was not palpable and no bruit could be heard. The optic discs were physiological, while acuity, colour, and fields of vision were within normal limits.

The eye in exophthalmos (Fig. 1b) was best studied with the patient lying on her right side and the head tilted slightly forward. It could also be evoked by throwing the head backward, or when artificial means were employed to increase intracranial pressure—for example, by pressing the ipsilateral jugular vein or on forced expiration with compression of the nostrils. Within a few seconds the eye would proptose to its maximum with congestion

FIG. 1a. *The eye in the normal position.*

FIG. 1b. *The eye in exophthalmos.*

of the sclera and rapid filling of a cluster of veins over the orbital ridge. The right palpebral fissure was narrowed ($R = 6$ mm; $L = 9$ mm) and the eyeball seemed to be displaced forward, downward, and outward, but eye movements were possible in all directions. The exophthalmos measured $R = 20$ mm; $L = 17$ mm. The pupil was of normal size and reacted promptly to light. There was no diplopia and the pulsation seen in the erect posture was visibly increased but again was not palpable and no bruit was heard. On assuming the erect posture, the exophthalmos would disappear as rapidly as it presented itself. The patient did not complain of any pain but within 5 min of proptosis was clearly uncomfortable.

FIG. 2. The radiograph of the skull shows erosion of the orbital roof.

FIG. 3. Right orbital phlebography.

Routine blood and urine examinations were normal. Blood serology was negative. The E.E.G. was unremarkable. The radiograph of skull and orbits (Fig. 2) revealed small areas of erosion of the right orbital roof.

Bilateral carotid angiograms (external and internal fillings) were inconclusive for any vascular malformations or tumours within the intracranial chamber or orbital cavities. A right orbital phlebography (Fig. 3) using the angular vein on the right side was carried out and showed (a) a cluster of vessels over the right supraorbital ridge, (b) a large vein crossing the left orbital cavity, presumably the left ophthalmic, and (c) numerous extracranial veins of normal size.

Under a presumptive diagnosis of intermittent exophthalmos, probably caused by orbital varices, the patient was subjected to a right fronto-temporal craniotomy. On elevating the right frontal lobe with the dura intact, the erosions in the anterior half of the orbital roof were seen and some of them extended through the depth of the bone, visualizing large, tortuous, and dilated veins within the orbit. The dura was next opened and surprisingly on the inner surface of its orbital portion was an area approximately $2\text{ cm} \times 1\frac{1}{2}\text{ cm}$ containing coils of vessels, knit together and closely adherent to the dura. These vessels were bright red in colour, pulsation in them was not clearly visible, but they were probably largely arterial coils conveying blood to the dilated veins within the orbit. No large artery was recognized. Being the main feeder of this dural arteriovenous malformation. The vessels were ligated and excised and a few were cauterized. The right orbit was unroofed and two or three large dilated veins forming part of the upper group were picked up, ligated, and excised. The cluster of subcutaneous vessels overlying the supraorbital ridge was similarly treated (Fig. 4).

The patient has been examined periodically since the operation, and a year later she reports that she has had no further attacks of exophthalmos nor can it be evoked by such means as were used before surgery.

DISCUSSION

The complete syndrome of intermittent exophthalmos is characterized by an alternating exophthalmos and enophthalmos and usually develops gradually and progressively. It has been seen at all ages, from babes in arms to those in the evening of life, but the great majority of cases present with symptoms and signs in the second and third decades. A case of bilateral involvement has yet to be seen and a survey of the literature shows a slight predilection for the left eye.

Vascular malformations within the orbit within it and the intracranial cavity have been found responsible for the disorder. According to Duke-Elder (1952) varicosities form 90% of the orbital anomalies, but arteriovenous aneurysms, hemangiomas, and lymphangiomas have also been encountered.

As regards symptomatology, the most common

FIG. 4. *The artist's impression of the lesion.*

complaint appears to be a bulging forward of the eyeball with a distressing feeling of fullness within it under conditions that would induce venous stasis within the affected orbit—for example, bending of the head forward or backward, excitement or straining, or even sudden changes of temperature as experienced by the patient in the case report. Eye pain is a complaint in only a minority of cases. The exophthalmos usually reaches its maximum within a few seconds and the patient is forced to seek relief by adopting such bodily positions to which he or she has become accustomed. In the exophthalmic state the patient's distress is obvious. The eyeball is pushed forward or downward and outward to as much as 30 mm or so. It may be congested and vision itself not infrequently reduced. Changes have also sometimes been observed in the pupil, which is dilated or narrowed. Eye movements may be restricted and diplopia may occur. Examination of the fundus may reveal slight congestion.

Walsh and Dandy (1944) observed pulsation of the eyeball in their patient and, in concluding dogmatically that an arterial component was present in the lesion concerned, proved it at operation. In our patient, faint pulsations were observed in the enophthalmic eye and these became more pronounced in the exophthalmic state. The weakness of the pulsations in our case was probably due

to the fact that they were transmitted through sieve-like openings in the orbital roof rather than through a widely dilated superior orbital fissure as was present in the case of Walsh and Dandy (1944).

In most of the reported cases, radiographs of skull and orbits were found to be negative, though the presence of an enlarged orbit or the superior orbital fissure has been noted in some cases. In an interesting review of the clinical material at the National Hospital and the Atkinson Morley in London, Rischbieth and Bull (1958) discussed the significance of enlargement of the superior orbital fissure and concluded with these remarks: 'The clue provided by widening of the superior orbital fissure in the diagnosis of cases of "orbital tumour" is valuable, since those lesions confined to the orbital cavity produce no bony erosion, whereas those with an intracranial extension show erosion in a high proportion of cases.' In our case there was erosion of the orbital roof.

Since attaining neurosurgical importance, many cases in recent years have had carotid angiography. This study has invariably been reported as normal, but even under these circumstances it would be fallacious to rule out the presence of an intracranial lesion, and especially if it be borne in mind that dural vascular malformations have been known to occur without the faintest trace of their presence on routine angiography (Obrador and Urquiza, 1952;

Verbiest, 1962) and which was so well exemplified in our own case. In fact the only clue to the presence of the intracranial lesion in this patient was the orbital erosion.

Krayenbühl discussed orbital phlebography in his valuable paper in 1962 and used the procedure himself in a case of intermittent exophthalmos. In a more recent publication, Arseni, Mihailescu, Simionescu, and Lê-Xuan-Trung (1965) reported their experiences in a series of 40 cases with orbital lesions of various types, and deduced some interesting conclusions from interpretation of the venous patterns seen on their radiographs. In our opinion, orbital phlebography in intermittent exophthalmos has one important value—that is, it helps to define the various communications that exist between the varices in the orbit and the intracranial chamber. It is known that each orbit has two sets of veins (upper and lower) which unite to form the main ophthalmic at the orbital apex and which eventually drains into the cavernous sinus. It is also known that the veins draining the lids, temporal region, and the face are in direct communication with these veins in the orbit of one or both sides, and, as in our case, are sometimes engorged. If therefore the venous stasis in the orbit in intermittent exophthalmos is influenced largely by changes in intracranial pressure, it would be reasonable to conclude that if only some of these dilated orbital veins were ligated at operation and/or even the main ophthalmic on the corresponding side were tied off, the remainder of the dilated veins in the orbit could still be influenced by changes in intracranial pressure through the ophthalmic vein of the opposite side, should a communication exist. In the case reported, orbital phlebography showed filling of a large vein, probably the ophthalmic in the contralateral orbit, a finding that could be of prognostic significance.

The syndrome, though self-limiting, is a progressive one and a retrobulbar haemorrhage is an occasional complication. Less frequently abducens paralysis, optic atrophy, and even sudden blindness have been reported.

The treatment of intermittent exophthalmos has been a lamentable tale of failures and complications. On the basis of pathogenesis it would seem clear that the aim of any operative procedure should be either (1) excision of all the offending veins in the orbit, or (2) interruption of these veins from their counterparts in the intracranial chamber. Most neurosurgeons and acknowledged ophthalmologists today would agree that the transcranial approach to the

orbit is the more acceptable, for it allows a wide exposure and a better definition of the lesions therein. Admittedly, at the time of this publication our patient has been followed up for too short a period to permit any operative conclusions. Nevertheless, it would seem to us on the basis of pathogenesis that, when an arteriovenous aneurysm is present in intermittent exophthalmos, it is imperative that the entire malformation including the orbital varices if present be excised as far as the limits of surgery will allow.

SUMMARY

A case of intermittent exophthalmos of the right eye occurring in a 41-year-old woman is reported. The cause was proven to be an intracranial arteriovenous aneurysm and orbital varices. Through a fronto-temporal approach, the anomalies concerned were extirpated with complete cure of the exophthalmos.

The literature on the subject has been reviewed and the syndrome discussed briefly with reference to its clinical features, radiological observations, and treatment.

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