UNILATERAL PROPTOSIS OF UNEXPLAINED ORIGIN*

PART III

BY

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ORBITAL exploration has shown that the commonest causes of unilateral proptosis of unexplained origin are small cavernous haemangiomas and blood cysts in the muscle cone space not extending to the orbital apex (Mortada, 1962a). When orbital exploration reveals no abnormality but is followed by slowly progressive proptosis, repeated skull x rays usually show that the cause is a deep orbital wall bony tumour or mucocele of an accessory nasal sinus (Mortada, 1963).

Further study of more cases of unilateral proptosis of unexplained origin has shown that the syndrome can be produced by other causes.

Case Reports

In the eight cases to be described the general condition of the patients was good. There was no important family history. The skull shape was normal and the face did not show any haemangioma or other abnormality. There were no signs of endocrine disturbances, no enlargement of lymph glands, or of the thyroid gland, liver, or spleen, or any palpable tumour in the body. Chest examination revealed no abnormality. There were no allergic manifestations, rheumatism, septic foci, or signs of vitamin deficiency. The skin did not show pigmentation or nodules. The nervous system; temperature, pulse, blood pressure, urine, and faeces were normal. The blood counts, total and differential, E.S.R., cholesterol, and bleeding and clotting times were normal. The blood Wassermann reaction and Casoni's test for hydatid were negative, and the tuberculin test was usually negative. Basal metabolic rate and ¹³¹iodine uptake tests were normal.

The ears, nose, throat, and nasopharynx were normal. Postero-anterior, oblique, lateral and base skull x rays and those of other parts of the skeleton were normal. There was no history of trauma and no acute inflammatory local signs such as oedema, redness of the lids, or conjunctival chemosis.

The proptosis was painless, not intermittent, not pulsating, forward in direction, and moderate in degree (about 19 to 22 mm.). Apart from the proptosis the eyes and their fundi were normal. There was no limitation of ocular movements, ptosis, unilateral myopia, or visual deterioration. No orbital mass could be palpated through the lids or conjunctival fornix, even after general anaesthesia.

* Received for publication December 29, 1964.

As the cause of the proptosis was unknown, the orbit was explored by lateral orbitotomy (Knapp, 1874; Reese, 1935).

Case 1, a 26-year-old male (Fig. 1), complained of right proptosis of 20 mm. (left side 16 mm.) of 9 months' duration. Orbital exploration showed in the muscle cone space below the optic nerve but not extending to the orbital apex a pink, soft 1×1 cm. encapsulated tumour. Histopathological examination of the tumour when removed showed delicate stellate cells lying in a homogeneous mucinous material (Fig. 2). The tumour mucin content stained red with mucicarmine. The picture was consistent with myxoma. After operation the proptosis was cured.

Case 2, a 29-year-old male (Fig. 3), complained of right proptosis of 22 mm. (left side 16 mm.) of 3 months' duration. Orbital exploration revealed in the muscle cone space below the optic nerve a tense 1×1 cm. cystic mass. During blunt little finger dissection, the cyst ruptured giving clear transparent fluid. The cyst wall was removed and found to be lined by one layer of endothelium. The lesion was diagnosed as a lymphatic cyst. One week after the operation the proptosis had disappeared (Fig. 4).

Case 3, a 15-year-old boy (Fig. 5), complained of left proptosis of 21 mm. (right side 15 mm.) of 7 months' duration. Orbital exploration showed in the muscle cone space, not extending to the orbital apex, a non-encapsulated hard grey 1×2 cm. mass which was adherent to the inferior rectus muscle. Biopsy showed a non-specific chronic inflammation with lymphocytic follicular aggregations (Fig. 6), consistent with a pseudo-tumour. Antibiotics and corticosteroids cured the proptosis.

FIG. 7.—(Case 4) Right proptosis in a woman aged 45 years. Orbital exploration showed reactive lymphocytic hyperplasia in muscle cone space.

FIG. 8.—(Case 4) Reactive lymphocytic hyperplasia. $\times 120$.

FIG. 1.—(Case 1) Left proptosis in a male aged 26 years. Orbital exploration revealed a myxoma in the muscle cone space.

FIG. 3.—(Case 2) Right proptosis in a male aged 29 years. Orbital exploration revealed a lymphatic cyst in the muscle cone space.

FIG. 4.—(Case 2) One week after removal of lymphatic cyst.

FIG. 5.—(Case 3) Left proptosis in a male aged 15 years. Orbital exploration showed a pseudo-tumour in the muscle cone space.

FIG. 2.—(Case 1) Myxoma, showing delicate stellate cells lying in a homogeneous mucinous material. $\times 120$.

FIG. 6.—(Case 3) Orbital pseudo-tumour showing non-specific chronic inflammation with lymphocytic follicular aggregations. $\times 120$.

FIG. 9.—(Case 5) Left proptosis in a woman aged 45 years. Orbital exploration showed increased size of the four rectus muscles due to idiopathic chronic orbital myositis.

FIG. 11.—(Case 6) Left proptosis in a male aged 31 years. Orbital exploration was negative. Thrombosis of left central retinal vein pointed to a diagnosis of idiopathic thrombosis of left orbital veins.

FIG. 12.—(Case 7) Right proptosis in a male aged 25 years. 5 years later a skull x ray and biopsy of right malar bone showed fibrous dysplasia of right orbital bones.

FIG. 13.—(Case 8) Left proptosis in a male aged 30 years.

FIG. 10.—(Case 5) Idiopathic chronic orbital myositis with lymphocytic infiltration of external rectus muscle fibres. $\times 540$.

FIG. 14.—(Case 8) Postero-anterior skull x ray taken 4 years later showing thicker and denser left lesser and greater wings of the sphenoid bone and a wider superior orbital fissure due to a meningioma of the left sphenoidal ridge.

Case 4, a 45-year-old woman (Fig. 7), complained of right proptosis of 19 mm. (left side 16 mm.) of 2 months' duration. Orbital exploration showed in the muscle cone space a non-encapsulated soft mass 1×1 cm. Histopathological examination of the mass after removal showed reactive lymphocytic hyperplasia (Fig. 8). The proptosis was cured by antibiotics and corticosteroids.

Case 5, a 45-year-old woman (Fig. 9), complained of left proptosis of 20 mm. (right side 16 mm.) of 3 months' duration. Orbital exploration showed that the four rectus muscles were thickened and bulky. Biopsy of the lateral rectus showed lymphocytic infiltration (Fig. 10), consistent with idiopathic chronic orbital myositis.

Case 6, a 31-year-old male (Fig. 11), complained of slight left proptosis of 19 mm. (right side 16 mm.) of 3 weeks' duration. Both ocular fundi were normal and the visual acuity in each eye was 6/6. While he was in the hospital for investigation he complained of diminution of vision of the left eye. Fundus examination revealed thrombosis of the left central retinal vein. Orbital exploration did not reveal any abnormality. The proptosis was diagnosed as due to idiopathic orbital vein thrombosis. Medical treatment with heparin and antibiotics cured both the proptosis and the thrombosis of the central retinal vein in 3 months.

Case 7, a 25-year-old male, complained of right proptosis of 19 mm. (left side 15 mm.) of 9 months' duration. Orbital exploration did not reveal the cause. The patient was seen again 5 years later (Fig. 12) when right proptosis had increased to 21 mm. There was right post-papilloedemic optic atrophy and the visual acuity had deteriorated to 1/60. There was prominence of right frontal and malar bones. Postero-anterior x ray of the skull showed the characteristic radiographic appearances of fibrous dysplasia of the orbital bones, areas of increased density being interspersed with porotic areas involving the lower parts of the right frontal bone, including the roof of the orbit, the greater wing of the sphenoid, malar, and upper part of the maxilla. The right lesser wing of the sphenoid also showed expansion and thickening and encroachment upon the superior orbital fissure. Oblique x ray showed a slight narrowing of the right optic canal. Lateral x ray showed thickening and expansion of the roof of the orbit extending backwards to but not involving the pituitary fossa. The other parts of the skeleton showed no abnormality. Again orbital exploration revealed no orbital tumour. A small portion of the surface of the affected malar bone was submitted for histopathological examination and the normal bone structure was found to have been replaced by delicate fibrous tissue characteristic of fibrous dysplasia of bone.

Case 8, a 30-year-old male (Fig. 13), complained of left proptosis of 19 mm. (right side 15 mm.) of 9 months' duration. Orbital exploration showed no abnormality. When the patient was seen again 4 years later, the left proptosis had increased to 22 mm. There was left optic atrophy with visual acuity of counting fingers at 0.5 m. Postero-anterior skull x ray (Fig. 14) showed thicker and denser lesser and greater wings of the left sphenoid bone. The left superior orbital fissure was wider than the right. Oblique skull x rays showed the left optic canal to be narrower than the right, suggestive of left sphenoidal ridge meningioma. Left orbital exploration showed irregular hyperostosis of the greater wing of the sphenoid. Transfrontal exploration by the neurosurgeon proved the diagnosis of left sphenoidal ridge meningioma.

Discussion

The subject of unilateral proptosis of unexplained origin and most of its causes is of extreme interest. Some of the aetiological lesions are rarely reported in ophthalmic literature.

It is uncommon to find orbital myxoma in the muscle cone space as described in the first case. Any lesion characterized by oedema and mucoid degeneration may simulate a myxoma (Blegvad, 1944). Five cases of myxoma of the orbit have been reported by Maucione (1914), Fuchs (1914), Lamb (1928), Bistis (1931), and Gifford (1931). The average age of the patients was 27 years, the oldest patient being 40 and the youngest 16.

Cystic lymphangioma of the orbit is rare (Jones, 1959; Mortada, 1962b). It is uncommon to find a lymphatic cyst in the muscle cone space as described in the second case. As regards orbital cysts containing clear fluid, Reese (1963) wrote: "Some authorities think that as an extreme rarity a serous orbital cyst may occur, which has its origin in the bursa either between the tendon of the superior oblique muscle and the trochlea or between the levator muscle of the upper eyelid and the superior rectus muscle." Also a serous cyst may develop in the sheath of a rectus muscle.

The presence in the muscle cone space of pseudo-tumour, as in the third case, or of reactive lymphocytic hyperplasia, as in the fourth case, and the occurrence of idiopathic thrombosis of the orbital veins, as in the fifth case, explain many cases of proptosis of dubious origin which regress by themselves or with medical treatment. Idiopathic thrombophlebitis of orbital veins simulating a primary orbital tumour is a rarity. Zimmerman and Rogers (1957) described a case in which during 17 days of observation the retinal veins became enlarged and tortuous without retinal haemorrhages or exudations.

Chronic orbital myositis may be due to syphilis, tuberculosis, orbital cellulitis, orbital phlebitis, or rheumatism. The rare idiopathic type of chronic orbital myositis as described in the sixth case has been reported by Gleason (1903), Offret (1939), Babel (1947), and François, Rabaey, and Evens (1956). Babel agreed with Offret that the idiopathic myositis was primarily a subacute or chronic endophlebitis of the orbital veins. As the pathology of the idiopathic condition is obscure, and the histological picture is not characteristic of inflammation but shows a patchy lymphocytic infiltration with fibrosis and degeneration of the muscle fibres resembling that seen in cases of endocrine disorder, Duke-Elder (1952) prefers to classify it with exophthalmic ophthalmoplegia due to thyroid pituitary disorder. If, during orbital exploration to discover the cause of exophthalmos, nothing is found, a biopsy of the rectus muscle should be taken to exclude dysthyroidism (Benedict, 1950).

Fibrous dysplasia localized to the orbital bones as described in the seventh case has been reported by Chartres (1953), Hobbs (1955), Matson (1958), and Mortada (1961). In its early stages the skull *x* ray usually shows no abnormality, and the proptosis is classified as of uncertain origin. Before diagnosing fibrous dysplasia of orbital bones, one has to exclude sphenoidal ridge meningioma and other orbital bony lesions giving a similar *x*-ray appearance. A biopsy of the affected bone must be examined histopathologically before the final diagnosis.

Sphenoidal ridge meningiomata are a common cause of unilateral proptosis (Elsberg, Hare, and Dyke, 1932). Skull *x* rays are helpful in 80 per cent. of such cases (Pfeiffer, 1943), but in others the changes in the lesser and greater wings of the sphenoid may not be clearly seen. When the lateral part of the lesser wing of the sphenoid is affected, the optic nerve is usually undisturbed for a long time. In this case the symptoms are cranial fullness with deformity in the temporal region and proptosis from involvement of orbital bones (Stender, 1933; Knapp, 1938; Smith, 1939). If the swelling of the temple is not apparent then all that eventuates is a unilateral exophthalmos. In these cases, if skull *x* ray and orbital exploration are negative as in the eighth case, the diagnosis is uncertain. Years may have to pass

before radiological changes or other signs and symptoms of sphenoidal ridge meningioma appear.

Summary

Unilateral proptosis of unexplained origin may be due to:

- (1) A small mass in the muscle cone space not extending to the orbital apex, as simple tumour, cyst, pseudo-tumour, or reactive lymphocytic hyperplasia. The commonest lesions are cavernous haemangioma and blood cyst.
- (2) Idiopathic chronic orbital myositis.
- (3) Idiopathic thrombosis of the orbital veins.
- (4) Early tumour or mucocele of an accessory nasal sinus.
- (5) Early deep orbital wall bony tumour such as fibrous dysplasia of the orbital bones or, in some cases, early lateral sphenoidal ridge meningiomata.
- (6) The described cases of orbital myxoma, idiopathic chronic orbital myositis, and idiopathic thrombosis of orbital veins are the first cases of such lesions to be reported from Egypt.

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