HAEMANGIOMA OF THE ORBIT*
REPORT OF A CASE

BY

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HAEMANGIOMATA frequently exist in the orbit for long periods before their nature can be determined. They are recognized as hamartomata, arising in congenital vascular abnormalities and enlarging slowly by canalization and increasing blood flow through their abnormal blood channels, rather than by true neoplasia. Haemorrhage from spontaneous or traumatic rupture, or thrombotic, cystic, or inflammatory episodes may interrupt this course, but in many cases they present simply as benign orbital tumours which, if situated posteriorly in the orbit, exert pressure upon the orbital contents. Among the familiar secondary effects of such pressure, loss of vision is, clearly, the most serious. This is emphasized by Walsh (1957) and was detailed by Birch-Hirschfeld (1930) in the well-known review of 92 cases, in 21 of which all vision had been lost in the affected eye and in 39 of which a more or less serious defect remained. Exposure keratitis and corneal ulceration from severe proptosis account for a certain number of such cases, and optic or retinal atrophy from direct pressure or following papilloedema is responsible for others, but there remains an appreciable proportion in which amblyopia is unexplained (Duke-Elder, 1952).

The following case is reported because the manner in which it presented is of interest; and because the visual loss, unassociated with objective evidence of a retinal or optic nerve lesion, proved recoverable after a period of 5 years' progressive increase.

Case Report

An educated man aged 48 was told by friends in 1953 that his left eyelid was sagging, and soon afterward became aware of impaired vision in this eye. Spectacles were prescribed and he was treated with orthoptic exercises for a time; but it was not until 1956 that he became aware of diplopia. Meanwhile the vision in the eye had further deteriorated, and he developed a vertical squint in addition to the apparent ptosis and began to experience temporal headaches, unaccompanied by nausea or vomiting. Latterly a suspicion of weakness of the left arm and vague paraesthesiae in the left thigh had resulted in neurological examination and, this being negative on the score of these symptoms, he was referred for further ophthalmological opinion.

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Examination (April 12, 1958).—The left eye was seen to be displaced upward some 8–10 mm. (Fig. 1), giving the appearance of ptosis, and some 4 mm. of proptosis was noted. The visual acuity in the right eye was 6/5, and in the left 6/36, corrected with −0.5 D sph., +3.5 D cyl., axis 75° to 6/18. Limitation of depression of the left eye was apparent; but vertical diplopia was elicited only on depression, left hyperphoria in the primary position being only 2 prism dioptres with the Maddox rod. The fundi showed no abnormality and no defect was found in the peripheral fields (¼° and 1° white objects). In the central fields the blind spots were of normal size and no scotomata were plotted.

Although no mass could be palpated in the lower part of the orbit, the ocular displacement, diplopia, and astigmatism were attributed to such a lesion, and the visual loss was thought to be due to the compression of the globe. No bruit was audible and skull x rays, whilst they raised the question of some expansion of the lateral wall of the orbit, revealed no abnormality of the optic foramina. Orbitonometry confirmed the degree of proptosis and indicated a degree of reduced repressibility suggestive of a solid mass within the orbit (Fig. 2).

The possibility that the tumour might be a haemangioma was considered at this stage and angiograms were undertaken; the angiogram revealed no abnormality but also produced no evidence of a retro-orbital mass. The phlebogram showed no abnormal
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intra-orbital venous spaces, no filling of the inferior orbital vein, and only a very narrow superior orbital vein displaced toward the orbital roof, with much reflux of dye down the facial vein. Figs 3 and 4 show the phlebogram of a normal subject compared with that of the patient.

Fig. 3.—Phlebogram of normal subject.

Fig. 4.—Phlebogram of patient, showing orbital flow through superior orbital vein only.
Operation (May 15, 1958).—Under general anaesthesia a mass was felt medially in the lower fornix, and its anterior end was exposed by a transconjunctival incision beneath the inferior rectus. The tumour was found to be easily separable by blunt dissection from the eye and extra-ocular muscles above and from the orbital floor below. It was lobulated and well encapsulated and was removed in toto after the division of a pair of small vessels. The incision was closed with a continuous silk suture, leaving a rubber drain which remained in situ for 5 days. Convalescence was uneventful.

Result.—Post-operative refraction (May 23, 1958) showed an immediate improvement in the visual acuity of the left eye to 6/9, which, when the astigmatism returned to its, presumably, normal level of 0-5 dioptre in 6 months, rose to 6/5. The normal appearance of the eyes was rapidly restored (Fig. 5), and the diplopia, evidently due to direct pressure upon the inferior rectus, disappeared.

Pathological Histology.—The tumour measured 28 x 23 x 14 mm. and was described as a sclerosing angioma composed of numerous thin-walled spaces containing blood. In addition, there were a few vessels with considerably thickened walls resembling arteries, but no elastic tissue could be demonstrated in them. There were a few collections of round cells in the interstitial tissue and also haemosiderin-laden histiocytes suggesting old haemorrhage. Scattered throughout the fibrous tissue were lymphoid follicles in which germinal centres were prominent.

Discussion

The post-operative recovery of vision in this case confirms the belief that visual loss arose directly from pressure on the globe which not only caused astigmatism, but also affected the retina itself. Such an effect is more familiar when oedema and retinal folds corresponding to the site of pressure are seen ophthalmoscopically and when, therefore, the probability of permanent reduction of visual acuity is greater than in the absence of such signs. That these do not necessarily indicate irreversible retinal changes, however, is demonstrated by the case recently reported by Hedges and Leopold (1959), in which the lesions were accompanied by pre-operative acuity of 1/60 improving to 6/9 after operation. Nevertheless, early diagnosis and treatment of the responsible tumour is clearly desirable, and to this end angiography, more especially phlebography, has a useful part to
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play. In this instance it was not possible to delineate the tumour radiographically, probably because of its scanty vascular connexions, although in other cases this has been possible (Boudet, 1954; Hobbs, DuBoulay, and Davis, 1960). On the other hand the indication of increased resistance to blood-flow through the normal venous channels of the orbit and the demonstration of upward displacement of the superior ophthalmic vein contributed materially to the diagnosis since such a finding is already familiar in cases of orbital tumour (Hobbs, Du Boulay, and Davis, 1960).

Offret (1951), believing that haemangiomata are common among benign orbital tumours, advised that when insidious unilateral proptosis was encountered this pathology should be the first to be considered, and to his advice may be added the suggestion that orbital angiography be then performed.

Summary

A case of unilateral ocular displacement caused by the pressure of a haemangioma, which also deformed the globe producing marked, temporary astigmatism and retinal compression, with full post-operative recovery of vision is described. The role of orbital angiography in the diagnosis of such lesions is briefly discussed.

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REFERENCES