CASE NOTES

ORBITAL ANGIOMA DIAGNOSED BY PHLEBOGRAPHY*

BY

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AMONG the causes of unilateral proptosis, tumours of the orbital contents must be frequently considered and, whilst their nature varies widely and published series differ considerably in their estimate of the frequency of different types of tumour, there is general agreement among ophthalmic surgeons that haemangiomas of various types are responsible for a large proportion of them. Thus, Reese (1941) found 25 of a series of 109 orbital tumours to be of this type, Forrest (1949) 25 of 184, and Drescher and Benedict (1950) seventeen in a total of 177. Duke-Elder (1952) refers to them as "one of the commoner orbital tumours".

Diagnosis of these tumours has usually rested on the clinical features of the orbital and ocular disturbances, assisted in some cases by the presence of other vascular abnormalities, commonly cutaneous haemangiomas. Such a diagnosis must perforce frequently remain presumptive for a long time, and exploration, if advised, is often delayed for various imperative reasons until to the initial deformity is added increasing pain and/or visual loss, which is commonly permanent.

Radiography in the Diagnosis of Orbital Tumours

Radiography has, until recent years, offered only limited help by demonstrating enlargement of the orbit or optic foramen or the presence of phleboliths, although the introduction of air or radio-opaque dyes into the orbital tissues has indicated, by a filling defect, the site of the lesion in some cases. Radiographic visualization of the orbital vessels has been recognized for some time as a possible further aid to diagnosis, either through the demonstration of displacement of these vessels or, more directly, by revealing a pooling of dye in the vessels of the tumour itself, thus both demonstrating its location and giving valuable information as to its nature. Attention was drawn to the potential value of arteriography employed in this way by Schurr (1951). Verified results from this method have been few, although Grino and Billet (1949) were able to demonstrate abnormal

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vessels in three tumours which were producing proptosis, two of which—a secondary hypernephroma and a meningioma—lay within the orbit. More commonly, however, positive results from arteriography have been confined to cases in which the abnormality was predominantly arterial, as in the orbital aneurysm reported by Huber (1951) and the angio-endothelioma found by Röttgen (1950). The commoner cavernous and capillary haemangiomas are not reported as having been demonstrated by this method possibly because, as is seen at operation, their arterial supply is often so slight. Our own experience in this respect illustrates this difficulty for, in three recent cases, although excellent delineation of the orbital arteries and filling of the choroidal circulation was seen, there was no outline of the arteries of the tumour subsequently removed at operation.

On the other hand opacification of the orbital veins by injection through the angular vein—orbital phlebography—appears to offer a more fruitful approach in some cases. By it the orbital venous pattern is readily visualized and abnormalities may be demonstrated either as displacement of the normal vessels or pooling of the dye in the tumour vessels. Haemangiomas which are largely of cavernous structure, as would be expected, lend themselves well to demonstration in this way, and in the case to be described the tumour was of this type.

Case Report

The patient was a woman, 20 years of age, in whom proptosis of the right eye had first become noticeable at the age of 4 years. Its onset after a cold suggested a rhinological origin and, although investigations revealed no positive pathology, sinus disease was accepted as its cause, and the condition ceased to attract attention until the patient was 14 years old, when the eye became slightly more prominent. She was first referred to one of us (H.E.H.) 2 years later, when the sole additional fact elicited was that a "strawberry mark" on the abdomen had been treated with CO₂ snow in infancy.

Examination.—The eye was displaced forwards, downwards, and outwards, with some swelling of the upper eyelid producing ptosis. A small collection of dilated lymph vessels was visible, subconjunctivally, on the temporal side of the globe. No other ocular abnormality was present, the retinae being of normal appearance, visual acuity excellent (6/5; 6/5), and binocular function unimpaired. No bruit was audible over the eye or orbit. Radiography of the orbit was negative, the orbits being of equal size. A provisional diagnosis was made of a tumour in the upper, inner quadrant of the orbit, probably of congenital origin and possibly a haemangioma; but exploration was not urged in view of the believed benignity of the condition. Subsequent observation over the next 4 years revealed little change, the proptosis increasing little, although later radiographs showed some enlargement of the orbit (Fig. 1, opposite). By this time, however, the patient was becoming increasingly dissatisfied with her appearance (Fig. 2, opposite), and pressed for further investigation and, if possible, treatment.

Angiography was, therefore, considered and carotid arteriography was performed. The vascular displacement revealed indicated simply a slight downward displacement of the globe. A phlebogram *via* the angular vein, however, revealed a large upper orbital vein breaking up into irregular, dilated branches just above the eyeball, and some of these branches were seen to run posteriorly into a dilatation in the postero-superior part of the orbit (Figs 3 and 4, opposite).

FIG. 1.—Radiograph of orbits at age 7.

FIG. 2.—Pre-operative appearance.

FIG. 3.—Phlebogram, antero-posterior projection.

FIG. 4.—Phlebogram, oblique projection.

Operation.—The nature and site of the tumour having been confirmed, its treatment was seen to be a neurosurgical problem, and it was removed at operation by Mr. Valentine Logue *via* an approach through the orbital roof between the superior rectus and superior oblique muscles. It is noteworthy that the tumour was found at operation to be supplied by a single small arterial branch.

Pathology.—Subsequent histological examination (Dr. W. H. McMenemy) showed the tumour to be an haemangioma approximating more to the cavernous than to the capillary type (Fig. 5).

FIG. 5.—Histological appearance of tumour.

Result.—The result has been excellent, both functionally and cosmetically (Fig. 6),* with but slight pulsation of the globe and a transient vertical diplopia which has entirely resolved.

FIG. 6.—Post-operative appearance.

Discussion

The salient feature of this case, and the reason for reporting it here, is the fact that the diagnosis, for long a presumptive one, was able to be made certain by orbital phlebography. This method of angioradiography would appear to have a field of usefulness in such cases as this which has so far been little explored in this country, although its value is becoming established on the Continent.

* This patient was demonstrated at a meeting of Clinical Section of the Royal Society of Medicine in November, 1958.

The technique originally described by Dejean and Boudet (1951) consisted in laying bare a length of the angular vein through a small incision; 20 ml. 35 per cent. Diodone solution were then injected through a fine needle into the lumen of the vein, and radiographs were taken immediately. In their paper and a subsequent one (Boudet, 1954), the authors give a detailed description of the radiographic anatomy in lateral and antero-posterior views, and mention the limitations of the method in visualizing veins in the inferior part of the orbit. They describe one case which appears from their illustrations to be similar to that which forms the subject of this paper, and, while discussing the investigation of vascular tumours, they point out the value of obtaining both an exact pathological diagnosis and an anatomical localization where a large varix can be demonstrated; they also suggest that the multiplication of opacified veins when these are dilated is a sign in favour of a vascular type of tumour.

Tumours of all sorts may reveal themselves by displacing the normal veins—particularly the superior orbital vein whose course is more or less constant. Boudet further suggests that some distinction between benign and malignant neoplasms may be possible from the radiological appearances. Tumours in the upper part of the orbit may not only cause displacement of veins, but even benign tumours may also so obstruct the flow that drainage is shown to be largely into the facial vein rather than into the cavernous sinus or the pterygoid plexus.

Yasargil (1957) introduced orbital phlebography in Zürich, and in some ways his technique is an improvement on that of Boudet, as he has been able, in a proportion of cases, to make his puncture of the angular vein directly through the skin, although he still finds it necessary to cut down on those veins not visible after jugular compression and dependence of the head. He stresses the importance of compression of any alternative pathways which the contrast medium might take (over the bridge of the nose and at the angle of the jaw), in order to obtain good filling of the orbital veins, and he uses only 8 to 10 ml. of contrast material. He also describes and illustrates two cases which are similar to ours but add little to Boudet's anatomical and pathological material.

Our own technique has been a purely percutaneous one, and except in a baby, we have not yet been forced to expose the angular vein. The area is infiltrated with local anaesthetic if the patient is considered fit to undergo the examination without general anaesthesia, and the angular vein is punctured blindly when it is invisible. The needle used is a fine one with a short bevel, and is attached to a length of fine polythene tubing. The needle and catheter are sufficiently light to remain in place without support, while clotting is prevented by the slow perfusion of saline. More than one injection (of 3 to 8 ml. of 45 per cent. Hypaque) is thus possible, and adjustments of position and obliteration of alternative pathways by pressure may be made.

The development of our own technique has so far been our chief exercise; but our findings in cases other than that one described in this paper have allowed us to confirm a number of Boudet's criteria. At the same time, we feel that the great variation in normal anatomy from case to case demands caution and experience in interpretation of the picture, particularly with regard to size of vessels and direction of flow.

We have also shown that a capillary haemangioma may not be accompanied by recognizable abnormality of the orbital veins; so that a negative orbital phlebogram following a normal carotid angiogram should still not be taken to exclude the presence of a vascular neoplasm.

How often the method may be expected to give such a positive result as was seen in this case is, of course, uncertain, and probably depends largely upon the size of the blood spaces within the tumour and the degree of communication which they have established with the general circulation. In large series of such tumours, such as that described by Ingalls (1953), the mixed capillary/cavernous structure seen in our case is common; and Ingalls's belief that the tumour starts in capillary form, the blood-containing spaces later enlarging to cavernous size, would appear reasonable, the lesion being, in fact, a form of hamartoma in which during life increased connexion with the circulation distends the congenitally formed cavities, as is known to occur in the cerebral angiomas. At a later stage, therefore, such a tumour would present as a simple varix with the more direct venous communications which readily transmit changes in venous pressure to it, and would result in the intermittent increases of exophthalmos with changes of posture or venous compression which are characteristic of orbital varices. Such a typical picture, whilst it may occur at any age, commonly appears only later in life, as Walsh and Dandy (1944) point out in their review of some 111 cases. Intermittent increase in the exophthalmos was not a feature of the case which we have described, but was produced in a slight degree with extreme jugular compression under anaesthesia, a fact which may be accounted for by the histological structure of the tumour. Nor was any bruit audible, a finding which is constant in the larger series of cases already mentioned.

It appears likely, therefore, that orbital phlebography offers a means of conclusive diagnosis in these cases at a stage earlier than is possible when their clinical features form the sole guide to their nature.

Summary

A case is described in which the cause of unilateral proptosis in a young woman was shown by orbital phlebography to be a vascular tumour, verified after removal as a cavernous/capillary haemangioma. The technique of phlebography is described, and its likely applications in such tumours is discussed.

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