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UNILATERAL PROPTOSIS OF UNEXPLAINED ORIGIN*

REPORT OF NINE CASES

BY

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REESE (1951) and other authors hold that, except when an orbital tumour is palpable, no surgery should be attempted unless a deep orbital tumour affects the function of the eye or the optic nerve. Reese recalled no case in which the orbit had been explored with benefit, merely because of an unexplained unilateral proptosis.

Among my series of 130 cases of unilateral proptosis, mostly caused by primary orbital tumours, there were nine in which the cause of the proptosis remained unknown. To leave such cases undiagnosed until an orbital tumour becomes palpable is a worry to both patient and ophthalmologist. The cause may be a malignant tumour, which may infiltrate the orbital tissues and give rise to metastasis. Even if the cause is a simple tumour or cyst, it is better to remove it while it is small and not yet firmly adherent to the orbital structures, and before it causes severe proptosis with the danger of corneal ulceration or presses on the optic nerve or ocular nerves causing visual deterioration and ophthalmoplegia.

Investigation of Nine Cases

The nine cases to be described occurred in patients whose ages ranged from 3½ to 60 years. There was no history of trauma or strain, and no acute inflammatory local signs, such as oedema, redness of the lids, or conjunctival chemosis. There was no subconjunctival haemorrhage or lid ecchymosis. The proptosis was painless, forward in direction, not intermittent, not pulsating, moderate in degree (19 to 22 mm.), of about 2 to 9 months' duration, and more or less stationary. Apart from the proptosis the eyes were normal. There was no limitation of ocular movements, ptosis, or unilateral myopia. The fundi were normal and there was no visual deterioration. No orbital mass could be palpated through the lids or conjunctival fornix, even after facial akinesia or general anaesthesia.

The general condition of the patients was good. 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 pre-auricular, submental, submaxillary, cervical, or other lymph glands, or of the thyroid gland, liver,

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spleen, and no other tumour in the body. Chest examination revealed no abnormality. There were no allergic manifestations or signs of vitamin deficiency. The skin did not show pigmentations or nodules. The ears, nose, throat, nasopharynx, and central nervous systems were normal, as were the temperature, pulse, blood pressure, urine, faeces, blood total and differential counts, sedimentation rate, and bleeding and clotting times. The blood Wassermann reaction and Casoni's test for hydatid cyst were negative, and the tuberculin test was usually negative. Basal metabolic rate and ^{131}iodine uptake tests were normal.

Postero-anterior skull *x* ray, with 20° tube tilt, showed no orbital dilatation, increased soft tissue density, or tumour calcification. There was no fibrous dysplasia of orbital bones, sphenoidal ridge meningioma, or any other orbital bony disease. There was no osteoma, mucocele, or malignant tumour of the nasal sinuses. The lesser and greater wings of the sphenoid, and the ethmoid, frontal, and maxillary sinuses were normal. There was no orbital bony destruction by a malignant tumour, and no bone destruction with the periosteal reaction of osteomyelitis. Oblique skull *x* rays did not show dilatation of the optic canals. Lateral skull *x* rays revealed no change in the sella turcica, clinoid processes, or sphenoid sinuses. In the series of 200 cases of exophthalmos reported by Pfeiffer (1943), roentgenography was diagnostic in 42 per cent., and suggestive in 20 per cent.

Exploration

As the cause of the proptosis was unknown, a deep orbital transconjunctival exploratory little finger palpation was performed as follows:

A lateral canthotomy to the external orbital bony margin is performed. The lateral fornix and Tenon's capsule are cut longitudinally (Knapp, 1874; Reese 1935). The lateral rectus muscle is pulled forward by a strabismus hook and the intermuscular fascia between the inferior and lateral recti is cut. The little finger is introduced into the fascial opening to feel the lateral, lower, and medial orbital walls, the posterior hemisphere of the globe, and the orbital part of the optic nerve. If no mass is felt, the intermuscular fascia between the lateral and superior recti is cut so that the little finger palpates the superor orbital wall, the posterior surface of the globe, and the optic nerve from another direction. The stretched oblique and rectus muscles must not be interpreted as tumours.

Dissection

If a tumour is present, its site, size, shape, consistency, and adhesions are noted, and it is removed by blunt dissection with the little finger.

Cysts usually rupture during their dissection, and care must be taken to avoid injury of the optic nerve, globe, and orbital muscles. Bleeding is not troublesome, and there is no need for post-operative orbital drainage. At the end of the operation, the conjunctiva and skin wounds are sutured.

This method of deep palpation and blunt dissection of orbital tumours is safe and has no complications. The nine cases are summarized in the Table (opposite), and the procedure is exemplified by Figs 1–4 (overleaf), showing details of Case 1.

A follow-up of these nine cases for 3 years has shown no evidence of recurrence of the proptosis.

TABLE

SUMMARY OF FINDINGS IN NINE CASES

Case No.	Age (yrs)	Sex	Side Affected Size (mm.)	Duration of Proptosis (mths)	Lesion in Orbital Muscle Cone Space not extending to Orbital Apex	Treatment	Prognosis
1	5	Male	R22	2 (stationary)	0·75 × 1·5 cm. cavernous haemangioma	Excision	Cure
2	10	Female	L19	2 (stationary)	0·5 × 1 cm. cavernous haemangioma	Excision	Cure
3	45	Male	R20	9 (stationary)	1 × 1·5 cm. fibrosing cavernous haemangioma	Excision	Cure
4	50	Female	R21	3 (stationary)	1 × 1 cm. fibrosing cavernous haemangioma	Excision	Cure
5	3·5	Male	R20	1 (gradually progressing)	Cavernous haemangioma with three blood cystic formations each 0·25 × 1·5 cm.	Excision	Cure
6	15	Male	R20	2 (gradually progressing)	Cavernous haemangioma with two blood cystic formations each 0·5 × 0·5 cm.	Excision	Cure
7	30	Female	R22	3 (gradually progressing)	1 × 1 cm. blood cyst	Puncture	Cure
8	14	Female	R19	5 (regressing)	Negative (probably absorbing encysted blood)	Drainage	Cure
9	23	Female	L21	4 (regressing)	Negative (probably absorbing encysted blood)	Drainage	Cure

Discussion

In seven out of the nine cases, orbital exploration showed that the lesion was in the muscle cone space, not extending to the orbital apex. Four cases were due to cavernous haemangiomata, two to cavernous haemangiomata with blood cystic formations, and one case to a blood cyst. In two cases the result of the exploration was negative.

The finding that in most cases the proptosis was due to a cavernous haemangioma is not surprising, since this is the commonest cause of unilateral proptosis due to a primary orbital tumour (Birch-Hirschfeld, 1930; Reese, 1941; Forrest, 1949; Duke-Elder, 1952). Its usual site in the orbit is the muscle cone space; it is a congenital tumour, slow-growing, and

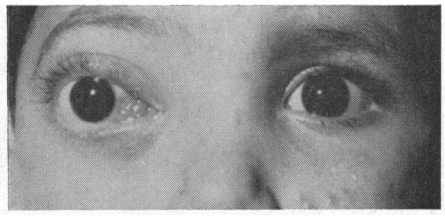

FIG. 1.—Case 1, right unexplained proptosis in a boy aged 5 years.

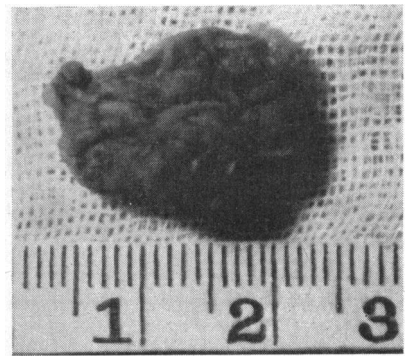

FIG. 2.—Case 1, small cavernous haemangioma found in the muscle cone space.

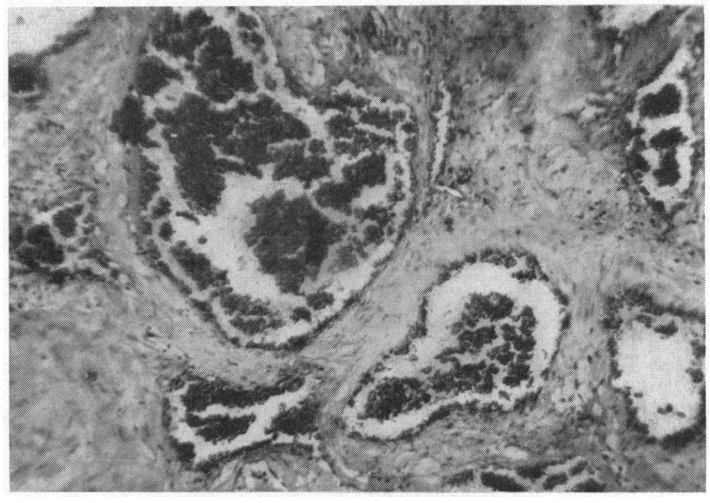

FIG. 3.—Case 1, cavernous haemangioma (× 120).

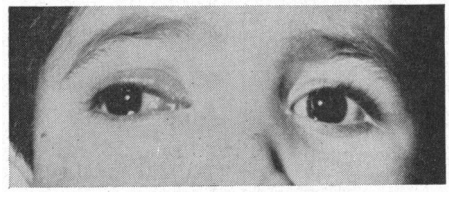

FIG. 4.—Case 1, showing disappearance of proptosis after excision of cavernous haemangioma.

occurring at any age. The motility of the globe remains unimpaired for a long time, and spontaneous sclerosis of the tumour may diminish its size (Gross and Wolbach, 1943).

Cases of cavernous haemangiomata with blood cyst formations may suggest a probable cause of orbital blood cysts and spontaneous haemorrhage of uncertain origin. Blood cysts may develop from the distension of spaces in an inconspicuous cavernous haemangioma, the lining endothelium

being destroyed by the pressure of the contained blood. They may also arise through haemorrhage in the tumour stroma. Spontaneous orbital haemorrhage may then be caused by the rupture of these blood cysts. Haemorrhage in the muscle cone space does not give rise to subconjunctival haemorrhage or lid ecchymosis, as the space is limited anteriorly by the union of the intermuscular membranes with Tenon's capsule (Charpy and Clermont, 1911).

If the blood cyst or haemorrhage is situated in the zone of the superior orbital fissure, a superior orbital fissure or orbital apex syndrome may develop. Such cases of proptosis or ophthalmoplegia usually recover spontaneously within 2 to 6 months by the absorption of the blood. Orbital blood cysts not lined by epithelium or endothelium resulting from the break-down of a spontaneous haematoma have been reported by Denig (1902), d'Amico (1924), Awerbach (1933), Wheeler (1937), and Svoboda (1948).

The cases in which the orbital exploratory findings were negative and in which the proptosis appeared to be cured, were probably due to encysted blood in the muscle cone space which drained away during the exploratory manoeuvres.

The rare cases in which orbital exploration is negative but is followed by progressive proptosis should be kept under observation. Further skull *x* rays and ocular and orbital examinations will probably determine when the orbital exploration should be repeated.

Summary

(1) The technique of orbital exploration described has no complications. It shows that the two commonest causes of unilateral proptosis of unexplained origin are small cavernous haemangiomata, and blood cysts in the muscle cone space not extending to the orbital apex.

(2) The origin of these orbital blood cysts and of spontaneous haemorrhage of uncertain aetiology in the muscle cone space is usually an inconspicuous cavernous haemangioma.

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