PRIMARY EXTRADURAL INTRA-ORBITAL MENINGIOMA
IN A CHINESE GIRL*

BY

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PRIMARY intra-orbital meningiomas are rather uncommon, and primary intra-orbital meningiomas unattached to the optic nerve sheath are decidedly rare. Scarpa (1816) was quoted by Byers (1901) as being the first to have reported an intra-orbital meningioma. Goar (1926) and Mayer (1928) estimated the number of intra-orbital meningiomas in the literature at 40, but there was no clear indication in many of these as to whether they were primary or secondary. Schreck (1939) studied 15 of these intra-orbital tumours removed at the Heidelberg University Hospital. Cushing and Eisenhardt (1938) saw only one in their series of 313 meningiomas. Craig and Gogela (1949) wrote a brilliant paper quoting 17 primary and 35 secondary intra-orbital meningiomas.

Primary intra-orbital meningiomas with no attachment to the optic sheath are rarer still and their pathogenesis poses a challenging problem. Only a few such tumours have been reported. Schreck (1939), in his Heidelberg studies, concluded that free-lying meningiomas need not arise from the optic sheath. Out of the 17 primary intra-orbital meningiomas of Craig and Gogela (1949), 3 were foraminal, 9 arose from the optic sheath, and 5 "lay more or less freely either within or outside the muscle cone or were attached firmly to the periorbita".

The case reported here is of a meningioma occurring outside the muscle cone and firmly attached to the periorbita of the medial wall of the orbit, and has a nerve coursing through it.

Case Report

A girl aged 9 years was admitted to the General Hospital, Singapore, on April 8, 1964, with a history of proptosis of the left eye for one year. The proptosis was gradually getting worse and was associated with headache but no vomiting. She had no diplopia.

On examination, the left eye was found to be markedly proptosed. Her general condition was good and systemic examination showed no abnormality. The central nervous system was normal and there was no abnormality of the cranial nerves.

She was referred to the ophthalmic unit for an opinion on April 9, 1964. Her visual acuity was normal 6/6 : 6/6. She had marked left unilateral proptosis which was directly forward for 8 mm. (compared with right). Movements were full, the proptosis was irreducible, the orbital margins were normal, the adnexa was normal, and the upper lid was normal in appearance and position with no lid lag. The anterior segment of the eye was normal, the pupils equal and reactive. The ocular media were clear and the fundus normal. There was no pulsation and no bruit. Further investigations were negative. The visual fields were normal.

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Skull X-ray.—The left optic foramen was normal. There was asymmetrical development of the paranasal sinuses, these being more prominent on the left than on the right. The left ethmoidal sinus encroached markedly on the left orbit, reducing the side-to-side diameter of its upper part. Some degree of unilateral proptosis would be expected from this cause alone, although this would naturally have been a long-standing condition.

Left Air Orbitogram.—The amount of air injected and retained within the left orbit was small and was mainly confined to the superior and anterior portion of the orbit, giving good definition of the upper border of the eyeball.

The left orbitogram was consistent with the presence of increased soft tissue in the left orbit in its medial and retrobulbar aspects. Its nature could not be established. It did not appear to show abnormal vascularity, as judged by the angiogram, which also failed to demonstrate any sizeable intracranial mass. The radiological evidence was considered to favour a lesion localized to the left orbit.

Left Carotid Arteriogram.—No abnormality shown. The ophthalmic branch of the left internal carotid artery could hardly be identified.

Operation.—The diagnosis of a retrobulbar space-occupying mass was made and it was decided to explore. At operation it was found that the retrobulbar space within the muscle cone was normal, but three well-circumscribed fleshy tumours were found on the medial wall of the orbit. These were found to be fixed and appeared to arise from the periorbita of the medial wall of the bony orbit. They were free from the bony orbit and separated from the medial rectus by orbital fat. Post-operatively, except for a wound infection which was controlled with antibiotics and temporary weakness of the lateral rectus and superior oblique, the patient made an uneventful recovery.

Pathological Findings

Gross.—Three pieces of tissue were obtained. The main tumour is about $15 \times 10 \times 5$ mm. and is roughly pyramidal in shape. It appears irregularly encapsulated. The cut surface of the main mass shows white and glistening tissue and some fat is included in it.

Microscopy.—The histology of the main tumour mass is that of a syncytial (meningothelio-matous) meningioma. The section has a base which is composed of vascular fibro-adipose tissue. The apex is rounded off and a small nerve is seen coursing over this tip just beneath the capsule (Figs 1 and 2). Below this there is some adipose tissue (retrobulbar fat) and the meningioma is seen infiltrating this fat (Fig. 3).

Fig. 1.—Note the irregularly encapsulated meningioma with the periorbita as its base. A small nerve trunk is seen curved along the top with retrobulbar fat beneath it. ($\times 3$.)

Fig. 2.—Higher magnification of nerve trunk, which is seen at the top of the picture. There is a collagenous band separating it from the meningioma below. ($\times 95$.)
The tumour cells (Fig. 4) themselves show a strongly eosinophilic cytoplasm. Rarely it is homogenous. There are no cellular outlines and the cytoplasm of one cell flows into that of another. These arachnoid cells are in a continuous sheet. The nucleus is mostly spheroidal and large, but sometimes it is oval or indented. A distinct nuclear membrane encloses a fine chromatin network in which one to three nucleoli are seen. No mitotic figures are present. Very occasionally a psammoma body is in evidence.

The general configuration of this tumour gives the impression that a more or less incomplete fibrous capsule tends to encircle the tumour. This in turn sends fibrous trabeculae into the tumour, and in some of these trabeculae nerve fibrils are seen.

The reticulin and collagen fibres are mainly confined to these trabeculae.

Comment

It is now generally agreed that syncytial (meningotheliomatous) meningiomas arise from the arachnoid cells of the meninges. Intracranially, the dura and periosteum are closely adherent, and to all intents and purposes they are one structure. However, on approaching the optic foramen they join the leptomeninges to start their journey along the optic nerve. The periosteum and dura part company once they are within the orbit. The dura accompanies the arachnoid and pia to cover the optic nerve up to the eyeball, and the outer layer of periosteum performs its normal function as the periorbita. Craig and Gogela (1949) took transverse vertical sections of the contents of the posterior two-thirds of the orbit at several necropsies. They could find clusters of arachnoid cells only along the meningeal coverings of the optic nerve. These cells were not found anywhere else in the orbit. The periorbita (like periosteum in other parts of the body) is composed only of dense collagenous fibrous tissue. Certainly this structure cannot conceivably give rise to a syncytial (meningotheliomatous) meningioma. The three possible ways of development are as follows: (1) occasional arachnoid "rests" in the periorbita could have given rise to these tumours; (2) all these extradural tumours were originally from the optic sheath and they had lost their connexion with the optic sheath by the time of discovery; (3) some smaller nerves are also endowed with arachnoid cells along their sheaths. Craig and Gogela (1949) tend to adhere to the second theory exclusively. However, we find it rather difficult to see how our tumour can possibly
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penetrate the layers of the muscle cone and lose its connexion with the optic nerve sheath and be strongly adherent to the periorbita after taking on a good layer of fibrous encapsulation. The fact that our tumour has a nerve trunk within it suggests the possibility that meningeal coverings with arachnoid cells do accompany other smaller nerves, besides the optic nerve. This particular nerve is probably a branch of the anterior ethmoid nerve.

Summary

A primary extradural, intra-orbital, syncytial (meningothelemiatous) meningioma is described. The pathogenesis of the tumour in this particular site is discussed, and it is suggested that this particular tumour arose from meningeal coverings of a tiny nerve outside the muscle cone coursing along the medial wall of the orbit, closely applied to the periorbita.

REFERENCES

SCARPA (1816). Cited by Byers.