MENINGIOMA INVADING THE OPTIC NERVE*

BY

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MENINGIOMATA of the optic nerve are rare tumours, although recently they appear to be somewhat less rare than is suggested in the earlier literature. Thus Mathewson (1930) could find only 33 reported cases, but Craig and Gogela (1949) were able to describe a single series of seventeen cases, nine arising within the orbit, three from the nerve within the optic foramen, and five apparently from other orbital structures. Reese (1951) found as many as 21 meningiomata (site not stated) in a series of 251 primary orbital tumours. Isolated cases are, therefore, hardly worthy of a separate report, but the one here described shows unusual and hitherto unrecorded features: the patient is the youngest yet reported, in only one other case was there a similar invasion of the optic nerve, and the number of ocular structures involved appears to be unique.

Case Report

A boy aged 3 was brought to the Benn Eye Hospital on November 14, 1955, as his mother thought that his right eye had an inward squint, and that he could not see properly with it.

Examination.—It was found that he did not follow light with his right eye, and that there was a fibrous-looking mass behind the lens nasally.

Subsequent examination under general anaesthesia confirmed that there was a large white mass involving the nasal half of the fundus and obscuring the optic disc. The remainder of the retina was detached and a retinoblastoma was considered to be the most likely diagnosis. Enucleation was recommended but parental consent for operation was not at first forthcoming. Eventually enucleation was undertaken on January 4, 1956.

No evidence of extra-ocular tumour tissue was noted at the time of operation.

Further Investigations.—After the pathological report on the enucleated eyeball, full medical and neurological examinations were carried out, including x rays of skull, chest, and abdomen, but no abnormality was found.

Treatment.—Mr. A. R. Taylor, neurosurgeon, saw the patient and reported as follows:

“It is rather a difficult decision whether to subject the child to major neurosurgical procedures at the moment. Reading the pathological report carefully one cannot be absolutely certain that the whole tumour has not been removed. In any event we are losing nothing by waiting a little, because if the tumour recurs, it will not infiltrate into the cerebrum; it will form a mass which will remain removable. By keeping an eye on the orbit and x-raying the optic foramina at regular intervals, I think that we will have early notice of any recurrence.”

No treatment other than enucleation of the right eyeball has been undertaken.

Present Condition.—He has remained well since and when seen in October, 1956, the right socket was healthy and no abnormality was noted in the media or fundus of the left eye.

X rays of the skull, including the optic foramina were normal.

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Observation.—It is proposed to repeat these examinations at regular intervals until we are reasonably certain that the tumour originated in the optic nerve and that it has been completely removed.

Pathological Findings.—The right eye was fixed in 10 per cent. formol saline.

Macroscopically the optic nerve measured 1.5 cm. from the globe to the cut end and showed no swelling. There was a funnel detachment of the retina with the inner layers in apposition, and on the naso-inferior side close to the disc the retina was swollen.

Microscopically there were two large masses of growth within the optic nerve close to the sclera immediately posterior to the lamina cribrosa. The larger mass had replaced the whole of the optic nerve at this point and had spread to the pia on one side, while the smaller mass surrounded the central retinal vessels and lay immediately posterior to the larger one (Figs 1 and 2).

FIG. 1.—Posterior half of globe showing retinal detachment and meningeomatous invasion of:

- (a) Inner layers of retina;
- (b) Choroid on temporal side of disc;
- (c) Optic nerve head;
- (d) Choroid on nasal side of disc and optic nerve.

Haematoxylin and eosin $\times 10$.

FIG. 2.—Opposing layers of detached retina invaded by meningeomatous growth. Haematoxylin and eosin $\times 90$.

FIG. 3.—High-power view of optic nerve - head showing typical whorls of meningioma. Haematoxylin and eosin $\times 390$.

Both were composed of densely-packed aggregations of large fusiform endothelial cells arranged in partial whorls segregated from each other by fine fibrous septa (Fig. 3). Tumour cells were present in the optic disc, in the adjacent choroid on both sides, and also in the stratum opticum of the detached retina. A transverse section of the cut end of the optic nerve showed no abnormality except for a collection in the extradural position of meningeomatous cells surrounding two psammoma bodies (Figs 4 and 5, overleaf).

FIG. 4.—Cross-section of cut end of optic nerve showing normal nerve and sheaths with focus of meningeomatous cells and psammoma bodies extradurally. Haematoxylin and eosin $\times 36$.

FIG. 5.—High-power view of extradural focus shown in Fig. 4. Note concentric laminated psammoma bodies. Haematoxylin and eosin $\times 195$.

Discussion

Meningiomata of the optic nerve usually occur after the first decade and more than half of them after the age of 30 (Coston, 1936). In a collection of 29 cases, Hudson (1912) found that more than 50 per cent. were over 30 years of age, and in none of the remainder did the onset of symptoms occur before

the age of 6. Forrest (1947) gathered together a collection of seventeen cases in which the average age was 40.

The age of our patient is therefore of interest; it is the youngest yet recorded although the case reported by Ryan (1953) may have been even younger when the lesion first developed. Ryan's patient was a boy of 6 at the time of operation, but by this age the growth had advanced to such a degree that the eye was blind and fixed in the orbit, and the neoplasm could not be completely eradicated. Moreover, there was a history of squint since birth.

In view of the youth of our patient, it is remarkable that so many structures had been invaded, *i.e.*, the optic nerve, central retinal vessels, optic disc, choroid, and retina, and yet the growth within the sheath of the nerve and outside it was minimal. Involvement of so many sites has not been reported before, although there are reports of invasion of one or two of these structures. In the series reported by Hudson (1912), invasion had occurred in the choroid in four cases, in the choroid and sclera in three cases, and in the optic nerve in one case. Coston (1936) believed that optic nerve meningiomata show a pronounced tendency to invade the globe, and he described a case of invasion of the optic disc and retina with the nerve remaining free, although, as is usual, it was compressed into a thin cord. One of the seventeen cases of Forrest (1949) showed growth extending along the central retinal vessels of the optic nerve, and in another the retina was invaded at the disc.

Neame (1923) reported a case in which a meningioma was present in the subdural space and also within the atrophic retina near the disc margin, but made no mention of invasion of the optic nerve. A somewhat similar distribution occurred in the case of a boy of 14 described by Hope-Robertson (1949), in which the subdural space of the nerve and the posterior aspect of the eyeball were involved.

We have found only one reference to definite invasion of the optic nerve fibres by meningioma cells. This was reported by Brailey (1887), who described a sarcoma growing from the dural sheath of the optic nerve and occupying the central artery and the nerve itself. A beautiful etching of the tumour shows it to be a meningioma.

Casari (1947) classified meningioma of the optic nerve into three types:

- (i) those growing from the sheath outwards,
- (ii) those infiltrating the sheath,
- (iii) those infiltrating the nerve.

However, he quoted no example of the third category, into which our case falls, and it is clear that, although invasion of the optic disc and retina may occur, extension into the optic nerve itself is extremely rare.

The problem of the exact site of origin of meningiomata has always excited controversy, and in the orbit, where a variety of differing structures

comes into close relationship, the question is particularly difficult. Indeed, it has been doubted whether meningiomata ever arise primarily within the orbit (Cushing and Eisenhart, 1938), and Verhoeff (1932), who on theoretical grounds accepted such an origin, had never seen a case. That they may do so, however, was well shown in a case of Friedenwald (1938), wherein a meningioma, together with the eye, was removed from the orbit; the proximal stump of the nerve adjacent to the foramen was normal, as also were x rays of this region, thus establishing the diagnosis of a primary intra-orbital meningioma.

The case here reported might be another such example, but the presence of tumour cells and psammoma bodies lying in the extradural region at the cut end of the optic nerve prevents a firm conclusion. It is possible that the tumour may have arisen intracranially or in the optic foramen, but there is no post-operative evidence of residual growth and the child is well.

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ADDENDUM

While this paper was in the press, a publication by Shepard N. Dunn and Frank B. Walse appeared in the *A.M.A. Archives of Ophthalmology*, Vol. 54, No. 5, p. 702, in which a case of meningioma invading the optic nerve is described.