Primary intraorbital meningioma

I. M. MACMICHAEL AND J. F. CULLEN

Department of Ophthalmology, Royal Infirmary, Edinburgh

Meningiomata comprise some 17 per cent. of all tumours of the central nervous system (Kernohan and Sayre, 1952; Reese, 1963). Of 230 consecutive cases of expanding lesions of the orbit, Reese (1963) found only eleven (5 per cent.) to be intraorbital meningiomata—none of them lying free within the orbit.

The large majority of these intraorbital meningiomata are secondary—that is they extend into the orbit from an intracranial site of origin (Van Buren, Poppen, and Horrax, 1957). Primary intraorbital meningiomata are rare and most of them have an attachment to the sheath of the optic nerve.

A comprehensive review was made by Craig and Gogela (1949), who collected a series of 35 cases of secondary orbital meningiomata and seventeen primary ones. Of the primary tumours nine were attached to the optic sheath, three arose from the optic foramen, and five were lying unattached to the sheath either within the muscle cone or outside it. These authors reviewed a few previously reported cases. More recently single cases of free-lying orbital meningiomata have been reported by D’Alena (1964) and Tan and Lim (1965).

The case reported here is of interest in two respects:

(1) It presented without the usual clinical features of visual defect or proptosis, so that the diagnosis was not suspected until histological sections had been examined.

(2) It was one of those primary meningiomata which at operation appeared to be unattached to other orbital structures.

Case report

A 20-year-old female student was seen in the Eye Department of the Royal Infirmary, Edinburgh, on October 16, 1967, complaining of a swelling in the right lower lid, associated with epiphora, which had been present for a little more than a year.

Examination An anterior orbital swelling was seen in the outer part of the right lower lid. The mass was firm and completely mobile and on its surface, which projected into the lower fornix, large vessels were visible (Fig. 1, overleaf).

Visual acuity was normal in each eye and there was no clinical proptosis, but it was possible to elicit diplopia on extreme downward gaze. X rays taken at this time of the orbits and skull were reported to be normal.

Progress The lesion was observed over a period of 6 months by which time it was larger and was causing diplopia in the primary position. Hess screen examination now showed limitation in the...
I. M. Macmichael and J. F. Cullen

I. Selling in right lower fornix with dilated conjunctival vessels running over it

The action of the inferior and lateral rectus muscles. A provisional diagnosis of haemangioma was made and it was decided to explore and remove the tumour.

Operation The tumour was approached through the skin of the right lower lid. The edge of the mass was well defined and it appeared to be encapsulated, shell ing out quite easily. It extended some way back towards the apex of the orbit but did not appear to have any attachment to the orbital tissues. After removal of the tumour, which now looked more like a dermoid cyst, some tissue posterior to it was excised for histological examination, to ensure that the tumour had, in fact, been removed in toto.

Pathological report (Dr. B. A. Bembridge)

"MACROSCOPIC: no obvious capsule is present but the surface of the specimen is slightly nodular, and gives the impression that it has been completely removed from the surrounding tissues.

"MICROSCOPIC: under low-power magnification, the appearance is of a sheet of cells predominantly spindle shaped. A capsule is present in some areas (Fig. 2). This is confirmed by higher magnification, and the cell nuclei are also seen to be long and narrow. There is a tendency toward whorl formation by these cells (Fig. 3). There are also occasional areas where the cells are more spherical, with correspondingly spheroidal nuclei. These tend to be nearer the periphery of the lesion, where blood vessels are also fairly frequent. A silver impregnation for reticulin shows a fair number of argyrophil fibres evenly distributed throughout the lesion. The histological appearance is that of a meningioma. No normal tissue is present to suggest a site of origin.

"The second specimen of tissue posterior to the tumour consists of collagenous tissue, areolar tissue, and blood.

"Diagnosis—MENINGIOMA OF ORBIT"

Result Postoperatively the visual acuity remained 6/6 and N5 in the right eye. Initially there was marked weakness of the right inferior oblique but this recovered rapidly.

Further x rays taken after the histological diagnosis confirmed the previous report of normal skull and orbits. Follow-up after 3 months showed no diplopia and no residual swelling.
Intraorbital meningioma

Fig. 2 Section of edge of tumour; a fibrous capsule is visible. Haematoxylin and eosin × 100

Fig. 3 Higher magnification demonstrating the tendency to whorl formation. Haematoxylin and eosin × 250

Comment

In previously reported cases of orbital meningioma the presenting features have included visual defect or proptosis or both. The case of Tan and Lim (1965) showed diplopia initially but subsequently developed proptosis. Clearly the tumour in these cases is lying far back in the orbit and either arises from or impinges upon the optic nerve.
In the case presented here the bulk of the tumour was lying far forward in the orbit giving rise to a swelling of the lower lid and a mechanical type of diplopia. In the series of Reese (1963) the commonest tumours found in the orbit were haemangiomata and malignant lymphomata. The possibility of a haemangioma was entertained in this case because of the large conjunctival vessels running over the surface of the tumour, and this was also statistically the most likely diagnosis. In this particular location dermoid cysts are not infrequent and at operation the consistency of the tumour suggested a dermoid cyst.

The only other report of a menigioma in an anterior site is that of Parzani (1933), who described a psammoma situated at the caruncle, extending into the medial part of the lower lid and bulbar conjunctiva, but the histology of his case was not typical of these tumours.

The most challenging problem of primary intraorbital meningiomata is their site of origin. Meningiomata in general are thought to arise from “cap” cells in arachnoid villi within the meninges (Boyd, 1961). The majority of primary intraorbital meningiomata, arising as they do from the optic nerve sheath, are therefore probably no exception in so far as there are arachnoid villi within the optic nerve sheath.

However, the exact site of origin of these meningiomata without apparent connexion to the meninges is less obvious. There are several possibilities:

1. The tumour was originally attached to the optic sheath but has migrated away from it (Craig and Gogela, 1949; Duke-Elder, 1952).

2. The tumour arises from arachnoid clusters of cells either along orbital nerves or in the interstitial tissues of the orbit as suggested by Tan and Lim (1965). Craig and Gogela (1949) were unable to find such cell clusters in serial sections of several fresh specimens obtained at necropsy of normal orbits.

3. The sutures between the orbital bones contain herniated meninges in a similar manner to that suggested by Kernohan and Sayre (1952) for extracranial meningiomata in the region of the glabella.

4. The tumour arises from the dura mater lining the orbit. The only two meningiomata of the fibroblastic type in Craig and Gogela’s series were both lying free within the orbit. Also the case reported by D’Alena (1964) and that reported here were both of the fibroblastic type. It seems possible that the fibroblastic type of meningioma at least may be derived from the periorbita.

Summary

A case of primary intraorbital meningioma is described and the site of origin of such tumours is discussed with an account of cases previously reported.

We are indebted to Dr. B. A. Bembridge who carried out the pathological examination in this case.

References

Intraorbital meningioma


D’ALENA, P. R. (1964) *Arch Ophthal. (Chicago)*, **71**, 832


PARZANI, C. (1933) *Policlinico (sez. prat.)*, **40**, 1492


Primary intraorbital meningioma.

I M Macmichael and J F Cullen

doi: 10.1136/bjo.53.3.169

Updated information and services can be found at:
http://bjo.bmj.com/content/53/3/169.citation

Email alerting service

These include:
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/