CASE NOTES

· ENDOTHELIOMA OF THE OPTIC NERVE*

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Case Report

An unmarried woman, aged 36, came to the out-patients' department with a history of having lost the sight of her left eye 3 years ago. The condition was diagnosed in another hospital as thrombosis of the central vein, and she was kept under 3-monthly observation. As far as she knew no aetiology of the trouble was ever found, and my direct inquiries at the source confirmed this.

Examination.—She was a fit woman at the time of her visit, with a blind left eye (no perception of light), and a typical enlarged amaurotic pupil. The right eye was normal. The left fundus showed a chalky white papilla with ? glial overgrowth at its lower half. The retinal vessels were seen to arch with this glial tissue and to dip to the fundus level at its border. All vessels—arteries as well as veins—were very thin, and the retina was pale.

There were no haemorrhages or exudates. This eye showed some limitation of upward and inward movement and it seemed to me proptosed, but these last two features were so indefinite that I asked the patient to bring me some old snapshots for comparison and, there was not much to go by even then (Fig. 1). She also had a history of tuberculosis.

FIG. 1.—Left proptosis exaggerated by utilizing the position of the eyes in which the patient's slight exophoria is best seen.

Diagnosis.—Tumour of the optic nerve, probably a glioma. From the clinical history it was deduced that the tumour started within the distal 10–15 mm. of the nerve, as occlusion of the central vein was the first sign of trouble.

Operation.—The proptosis was very indefinite and the x ray of the optic canal was normal I delayed surgery, but after 3 months of observation she was admitted to hospital for

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enucleation. This was started by lateral canthotomy and about 20 mm. of the optic nerve was removed with the globe (Fig. 2). Because of the very anterior location of the tumour I hoped to remove it in this way, without further mutilating surgery, but the section of the distal end of the nerve showed neoplastic infiltration.

FIG. 2.—Enucleated globe, showing thickened optic nerve, primary swelling just behind the eyeball, and second swelling distally.

The patient was referred to the neurosurgical unit at the Midland Centre for Neurosurgery for transfrontal approach, and a second operation was performed by Mr. R. C. Connolly. The optic nerve at the orbital extremity of the optic canal was found to be normal, and was divided at that point. The tumour, which involved the nerve immediately distal to that point and was densely adherent to the surrounding tissues of the orbit, was eventually removed *in toto*.

Microscopic examination of both specimens proved the tumour to be an endothelioma (Fig. 3 and Fig. 4, opposite).

FIG. 3.—Section through globe. $\times 2$.

Comment

This tumour, endothelioma of the optic nerve, is not common. Duke-Elder (1940) traced 33 described cases up to 1933, and all subsequently recorded cases would still not double that figure. In putting this case on record, two problems seem to be worth emphasizing: the diagnosis, and the nature of the proptosis.

The clinical diagnosis in this case presented no difficulties, but the pathological diagnosis depended entirely on the pathologist and the microscope. This tumour had all the clinical differential characteristics of a glioma: the comparative youth of the patient, the good mobility of the eye, early

FIG. 4.—High-power magnification of section of tumour, showing typical structure endothelioma.

loss of vision with vascular occlusion, no cranial extension, and no pain, but proved histologically to be an endothelioma. A similar case was described by Stallard (1935) with close retrobulbar localization of the tumour.

Considering only the increase in the bulk of the optic nerve, it seems highly improbable that proptosis is caused in these cases simply by the space-taking of the tumour within a rigid orbit. When I operated I felt that the proptosis was caused by the thickened and unyielding stalk of the optic nerve pushing the globe forward. Because of this thickening the nerve had lost its physiological curves, thus elongating and acting like a stiff rod. Otherwise the tissues of the orbit were lax and freely admitted a finger to palpate the tumour without any further forward displacement of the globe.

It is now about 2 years since the operation was done and the patient has remained well.

REFERENCES

- DUKE-ELDER, S. (1940). "Text-book of Ophthalmology", vol. 3, p. 3089. Kimpton, London.
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