DIKTYOMA

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TUMOURS of the ciliary epithelium are so rare, that probably not more than twenty cases of these neoplasms have been recorded. Their embryonic nature was first recognized by Verhoeff (1904), but the name diktyoma was coined by Fuchs (1908) for neoplasms of the ciliary epithelium in children. The general impression is that these diktyomata are only locally invasive and relatively benign, but Klien (1939) and Imre (1941) have each reported a case, emphasizing their malignant nature, and Malone (1955) reported a case showing extensive intracranial invasion on *post-mortem* examination. Reese (1956) discussed the macroscopic and microscopic appearance of the tumour in great detail and laid down definite criteria for histological diagnosis.

Case Report

A girl aged 6 years was seen in the Ophthalmic Out-patients Department on November 3, 1958. The father had brought the child with the complaint of a progressively increasing white reflex in the right pupil of one months' duration. There was no other history.

Examination.

Left Eye.—Quite normal with a visual acuity of 6/6.

Right Eye.—The visual acuity was 6/60. Cornea bright. Anterior chamber normal. Pupil dilated and sluggish. There was a white cystic mass on the temporal side, just behind the lens, extending from the 10 o'clock to the 4 o'clock position, which occluded almost half the pupillary aperture. Large numbers of blood vessels were seen on the surface of the mass. The fundus, as far as could be seen, was normal. The ocular tension was slightly lowered. The pre-auricular gland was not enlarged. X-rays of the orbit and chest showed nothing abnormal. Transillumination was negative.

Provisional Diagnosis.—Glioma retinae.

Treatment.—A diathermy puncture of the cystic mass was done on November 20, 1958, 8 mm. outside the limbus on the temporal side. The cyst partially collapsed and some fluid came out under the conjunctiva. A plastic iridocyclitis set in after the puncture, with exudate in the vitreous and the anterior chamber.

The eye was enucleated on November 29, 1958. When the eye was cut open, a greyish white mass about 10 mm. across was seen in the ciliary region with a thin membrane covering it. The case was followed up for 4 months after the enucleation.

Pathological Report.—The specimen was a bisected eyeball which on reconstruction measured approximately 21 mm. in sagittal diameter, 20 mm. in frontal diameter, and 18 mm. in vertical diameter. The cornea appeared to be slightly opaque. The lens had fallen out. Arising from the ciliary region was an irregular greyish-white firm tumour mass roughly 7 mm. in diameter, which extended posteriorly to a detached fold of the

retina. There were no areas of haemorrhage or necrosis, and the tumour tissue had not penetrated the other coats of the eyeball at any point.

The eyeball was fixed in Bouin's fixative, and sections stained with haematoxylin and eosin, Masson's trichrome, and Mallory's phosphotungstic acid and haematoxylin were studied. The tumour appeared to arise from the ciliary body pushing the iris in front of it. It consisted of tubules and bands composed of several rows of cells with prominent nuclei, lying perpendicular to their long axes. In some of the tubules, a delicate membrane resembling the external limiting membrane of the retina could be made out. In other parts, the tumour cells occurred in solid conglomerations without any rosette formation. The tumour cells did not contain pigment. The other outstanding components of the tumour were the fairly large islets of cartilage with partial calcification (Fig. 1). Mallory's stain showed nerve fibrils (Fig. 2).

FIG. 1.—Tumour cells arranged in tubules and bands and a strip of cartilage partly ossified (low power).

FIG. 2.—Neuroglial tissue (low power).

There was a complete absence of organized stroma. However, a framework of connective tissue containing a few thin-walled blood vessels was present between the tubules, cell aggregates, and islets of cartilage. The pigmented cells at the periphery of the tumour appeared to have been derived from the iris.

Summary

A case is reported of a diktyoma in a 6-year-old child.

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