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## CASE OF NEUROFIBROMA OF THE CHOROID\*

BY

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NEUROFIBROMA of the choroid is an extremely rare disease; so that any instance of it deserves reporting; recently a case has reached Moorfields, which, in many respects, is unique among the few on record.

*Case History.*—M. C., a girl aged 3½ years, had been noticed for the previous 4 months to have an increasing prominence of the left eye, being well in every other respect. She was then shown to her doctor, who referred her directly to Moorfields, and the following signs were evident:—The right eye was normal. The left eye was moderately proptosed, with some limitation of all movements, especially on looking upwards. There was no other abnormality save on ophthalmoscopic examination, which revealed a globular mass almost filling the supero-temporal quadrant, whiter than the adjacent choroid, but with distinct choroidal blood-vessels on its surface, and normal retinal blood-vessels directly overlying it; there was no pigmentary proliferation. The mass was sharply demarcated, almost reaching the optic disc; at the lower border was a trace of fluid detachment; elsewhere the fundus was normal.

The appearance was thus similar to that of a choroidal sarcoma with extra-ocular extension, but the age of the patient prevented this diagnosis.

X-ray and general examination showed no evidence of metastases or other abnormalities.

On May 14, 1943, Mr. Doyne performed an exploration of the

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\* Shown at the Section of Ophthalmology, Royal Society of Medicine, November 12, 1943.

left orbit, through division of the conjunctiva between the superior and external recti, and exposed a firm, circumscribed tumour, extending back from the sclera towards the apex of the orbit, transillumination showing the detachment to be opaque, exenteration of the orbit was effected forthwith. The socket healed well and has given her no trouble since then.

*Pathology.*—I am indebted to Mr. Goldsmith for permission to retail his report on the pathology of the tumour.

“The specimen consisted of the left orbital contents. The orbital fat and muscles were removed, exposing the left globe with a soft whitish mass adherent to it in the region of the insertion of the superior oblique muscle. This mass was irregularly lobulated and approximately equal in size to the globe itself. It had displaced the optic nerve downwards and inwards, and had caused considerable distortion and indentation of the whole posterior part of the eye. It was apparently encapsulated, and not invading the orbital tissues.

“The eye and mass were divided in a slightly oblique plane passing downwards and inwards. As is well shown in the illustration the mass was now seen to be composed of a smaller smooth rounded intra-ocular portion in the choroid, communicating through a fairly wide scleral perforation with a much larger irregular lobulated extra-ocular extension. The cut surface of the mass was yellowish in

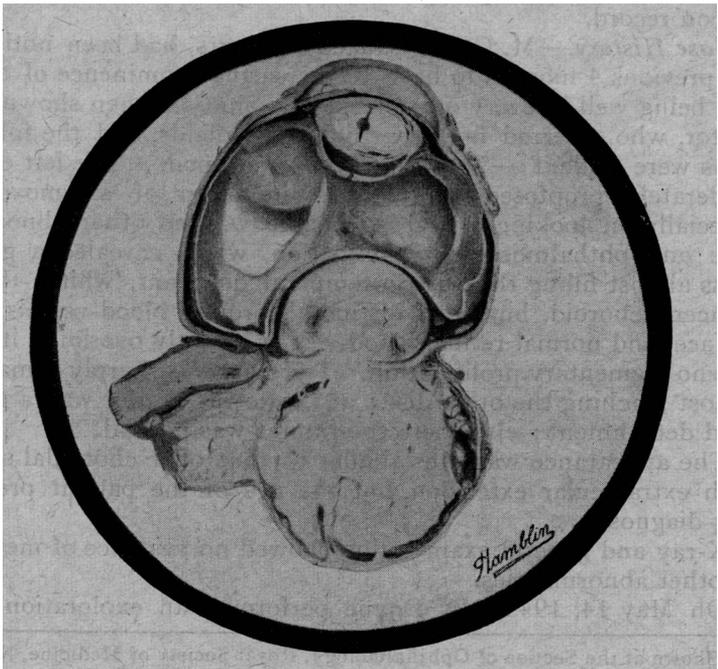


FIG. 1.

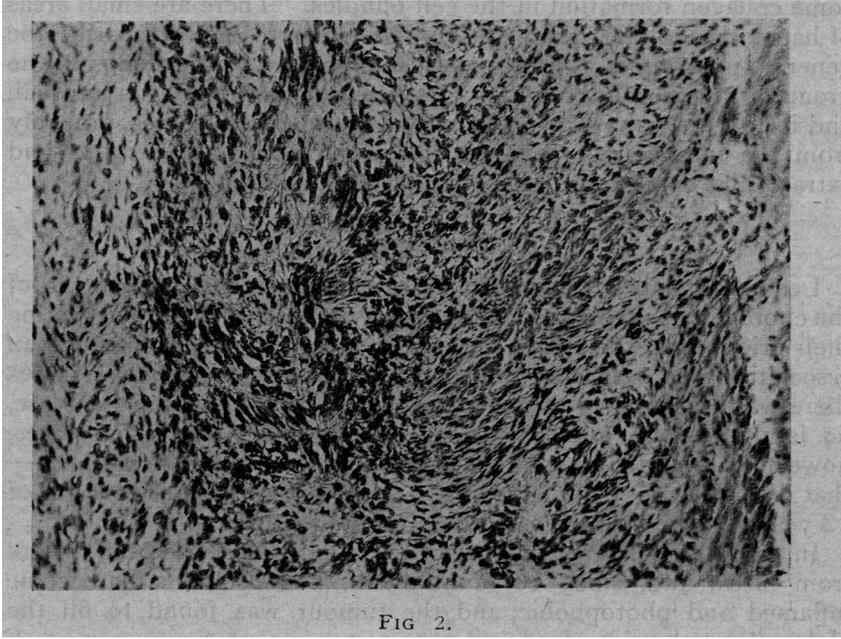


FIG 2.

colour and fairly soft in consistency, although crossed in places by firmer trabeculae of fibrous tissue. Half of the bisected specimen was embedded in celloidin, and sections were cut and stained with haematoxylin and eosin, and with haematoxylin and van Gieson's stain."

### Histology

The anterior segment of the eye is normal. To the general architectural features as seen with the naked eye and loupe and noted above, the microscope adds the facts that the retina, although raised over the intra-ocular part of the mass, is otherwise entirely unaffected. The tumour also has not invaded the constituent elements of the choroid, which is stretched out over its surface, the lamina vitrea being intact. A fairly large scleral perforation is present and the scleral edges have caused a constriction in the mass. The sclera is not invaded and its margins appear to have been separated by direct pressure.

The tumour itself has a well defined fibrous capsule, although this is poorly developed over the intra-ocular portion. It is composed of elongated cells showing well marked parallel arrangement, and many areas of palisading. It is unusually cellular, and there are a few mitoses. Collagen formation is correspondingly scanty but well developed fibrous septa can be seen, and there is

some collagen formation in the cell bundles. There are small areas of haemorrhage and degeneration. In spite of the cellularity the general appearances do not suggest malignancy. It belongs to the group of perineural fibromata. Its situation in the orbit is unusual, and it appears to have arisen from one of the ciliary nerves, probably from the intra-scleral portion; this would explain the intra- and extra-ocular extensions of a non-invasive tumour.

### Discussion

I can find only six recorded cases of discrete neurofibromata of the choroid; as in this case the majority are in a situation suggesting their origin from the ciliary nerve, although in one of them such an association was sought in vain. In common with the other cases there was no evidence of generalised von Recklinghausen's disease, no family history, and, as yet, no recurrences. The other cases, however are virtually confined to adults, the youngest instance—that of Kyrieleis—commencing as a small limbal tumour in a girl of 12 years. Two other cases show unusual features:—

In that of Papoleczy, a man aged 43 years, the eye had been shrunk from birth, and, since 18 months old, had occasionally become inflamed and photophobic, and the tumour was found to fill the globe, about the size of a hazel nut.

And in that of Callender and Thigpen, two tumours, the size of a pea, were present in the anterior and posterior regions of the same eye.

In contrast to these, a diffuse neurofibromatosis of the choroid occurs more frequently, usually in infants and resulting in glaucoma, and is usually associated with evidences of von Recklinghausen's disease elsewhere.

I am indebted to Mr. Doyne for permission to report this case, and to the Laboratory at Westminster Hospital for preparation of the microphotograph.

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