Choroidal neurilemmoma—an unusual clinical misdiagnosis

R. B. S. PACKARD AND J. HARRY

From the Institute of Ophthalmology, University of London

SUMMARY A case of choroidal neurilemmoma is reported. After a period of clinical observation which included serial fundus photography and fluorescein angiography, during which time active growth was seen, the tumour was thought to be a malignant melanoma, and enucleation was considered the treatment of choice. Subsequent histological study revealed a neurilemmoma. Important points relating to the clinical diagnosis of lesions presenting in this way are discussed.

Ocular neurogenic tumours in the absence of generalised neurofibromatosis are rare and only sporadic reports have appeared in the world literature¹⁻⁸. A neurilemmoma of the ciliary body was described by Donovan⁹ in 1956, and in recent years there have been 2 further reports of uveal neurilemmomas, one from Brewitt *et al.*¹⁰ in Germany and the other from Harada *et al.*¹¹ in Japan. An indication of the frequency of this particular growth is that Ferry,¹² in his large series of 7877 eyes, reported only 1 case.

Case history

A 43-year-old Caucasian woman presented on 4 November 1976 with the symptom of 3 months of increasing blurring of vision on the nasal side of fixation. She did not complain of either flashing lights or floaters, and apart from myopia there was no past ocular history. The patient had, however, suffered from hypertension for some years and was taking oxprenolol (Trasicor) and cyclopenthiiazide with potassium chloride (Navidrex-K).

On examination the corrected vision in the right eye was 6/24 and in the left 6/6, the anterior segments were normal, and the intraocular pressures were not raised. The only abnormality was in the right fundus, where an elevated greyish mass was seen supero-temporal to the macula (Fig. 1). The differential diagnosis at this time was between malignant melanoma of the choroid and disciform degeneration.

Fluorescein angiography 2 weeks later, although not diagnostic (Fig. 2), raised other possibilities,

Fig. 1 Fundal photograph showing raised subretinal mass.

and the differential diagnosis now lay between a haemangioma, a malignant melanoma, and a metastatic tumour. With the latter in mind the patient was fully investigated, but nothing of note was found.

For a time colour photography and fluorescein angiography were repeated regularly, and, in order to try to lessen an exudate which was threatening the macula, argon laser treatment was given. When the patient was seen 6 weeks later, however, the right visual acuity had dropped to 6/60, the tumour had increased in size, and a vascular pattern within the neoplasm was detected. Blood was taken to examine for melanoma antibodies, and this investi-

Correspondence to Dr J. Harry, Department of Pathology, Institute of Ophthalmology, Judd Street, London WC1H 9QS.

gation subsequently proved to be positive. Since the consensus of several ophthalmologists was that the tumour was a malignant melanoma, the eye was enucleated a month later. At operation no extra-ocular extension was seen.

HISTOPATHOLOGICAL REPORT

Sections showed a benign choroidal tumour arising posteriorly (Fig. 3) and composed of rather small spindle type cells showing palisading. Some conspicuous blood vessels were present, and the histological features were those of a neurilemmoma (Fig. 4). There was marked degeneration in the

overlying retina, and the cornea showed guttate excrescences on its posterior surface similar to those seen in Fuchs's dystrophy, but apart from these changes the remaining ocular structures showed no obvious significant gross abnormality.

Discussion

With the increasing use of indirect ophthalmoscopy, fundus photography, fluorescein angiography, and ultrasonography, the incidence of enucleating eyes containing tumours other than malignant melanoma should be diminishing. Two series of eyes enucleated

Fig. 2 Fluorescein angiogram showing abnormal vasculature in tumour mass.

Fig. 3 Section of the whole eye showing tumour in posterior choroid. (Haematoxylin and eosin, $\times 2.25$).

Fig. 4 Tumour spindle cells showing palisading. (Haematoxylin and eosin, $\times 350$).

for malignant melanoma and analysed by Ferry¹² and Shields and Zimmerman¹³ revealed that 19% and 20% respectively contained a simulating lesion. In other series, however, the incidence was reported as 5.6%¹⁴ and 8%.¹⁵ Harry¹⁶ had found an incidence of 11.3% in cases where a request had been made to exclude malignant melanoma. In 1974 Shields and McDonald,¹⁷ in analysing figures where modern diagnostic methods had been used, showed an incidence of misdiagnosis of 3.7% over 11 years. It was found that during the last 6 years of this study the incidence was 1.9%, this figure representing only 2 patients, both of whom had a metastatic carcinoma. These authors emphasised the importance of clinical awareness of such simulating lesions and also recognised that transillumination was useful as a diagnostic procedure. Reference is also made to the value of radioactive phosphorous uptake.

In the case under discussion, as can be seen from the history, a definitive diagnosis was not made initially because the tumour was atypical in shape and the fluorescein angiography had not provided a characteristic appearance. Therefore clinical observation, serial fundus photography, and fluorescein angiography were continued, and only when definite signs of growth and neovascular patterns within the tumour were seen was it decided that malignant melanoma was the probable diagnosis.

Gass,¹⁸ in outlining the problems of distinguishing benign from malignant uveal tumours, presented evidence that even observed growth was not a reliable sign of malignancy, but stated that if it did occur enucleation was probably the safest course of action. The case described illustrates how such growth may lead to misdiagnosis and removal of the eye. It is of interest that melanoma cytoplasmic antibodies were present, thus confirming the suggestion of Garner¹⁹ that immunological testing in such cases is inaccurate.

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