Bilateral primary choroidal melanoma

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SUMMARY A case of bilateral primary choroidal melanoma is described in a 51-year-old white male. Histological confirmation followed enucleation of the left eye and local excision in the right. A careful systemic evaluation failed to demonstrate any other primary or secondary melanomata. This case is the first bilateral primary choroidal melanoma reported in the United Kingdom and the first to be described in which histological confirmation was obtained as a result of local excision of one of the tumours.

The occurrence of choroidal melanomata as separate primary tumours in the 2 eyes of an individual is an extremely rare event. It has been estimated that, out of a population of 50 million whites, only one person will develop bilateral choroidal melanomata during his or her lifetime.¹

There are reports of cases of multiple foci of melanoma in one eye^{2–4} and of iris melanoma in one eye with choroidal melanoma in the second,^{5–7} but only 8 well documented cases of bilateral choroidal melanoma have been reported^{8–12} from various parts of the world. No case has yet been described in the United Kingdom.

Case report

A 51-year-old white man born on 14 June 1928 was initially referred to the Oncology Clinic in July 1979 after a 3-month history of blurred vision in his left eye. The referring ophthalmologist had noted a pigmented mass at the posterior pole of the left fundus. At the initial examination visual acuities were noted to be 6/6 and 6/12 in the right and left eye respectively. The anterior segments appeared normal. No abnormalities were reported in the right fundus. Examination of the left fundus, however, showed a solid pigmented choroidal mass, approximately 6×8×2.5 mm in size (Fig. 1) temporal to the macula. Orange spots were present on the surface of the lesion, and there was a small associated serous retinal detachment. Fluorescein angiography supported the diagnosis of choroidal melanoma, and the left eye was subsequently enucleated. Histology confirmed

the presence of a lightly pigmented spindle cell choroidal melanoma (Fig. 2). No evidence of extraocular extension was present.

The patient remained asymptomatic until early in 1981, when he noticed intermittent shimmering in the upper field of vision of his right eye. The visual acuity remained unchanged and his general health was good.

He attended the clinic in April 1981, when examination of the right eye revealed a pale solid tumour, approximately 8×8×10 mm, arising inferiorly from the anterior choroid and involving the pars plana (Fig. 3). The associated serous retinal detachment

Fig. 1 Left eye. Choroidal melanoma with overlying orange spots and associated serous retinal detachment.

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Fig. 2 Left eye. Lightly-pigmented spindle cell choroidal melanoma. ($\times 250$).

extended as far as the inferotemporal arcade of vessels. The margin of the detachment was demarcated by a line of fine pigment deposition. Other than slight narrowing of the drainage angle inferiorly no other ocular abnormalities were detected, and the intraocular pressure was 17 mmHg. The right visual acuity remained 6/6.

Fluorescein angiography highlighted the vascularity of the tumour (Fig. 4). There was leakage from these

vessels in the later stages of the angiogram. Ultrasonography showed features compatible with a melanoma. Systemic examination revealed no evidence of primary pigmented skin lesion, hepatomegaly, or metastatic disease.

A diagnosis of an anterior choroidal melanoma was made, and it was elected to manage the patient conservatively at that stage. Regular follow-up at 3-monthly intervals showed little discernible change in the size of the mass itself. However, the associated serous detachment increased slightly to encompass the inferior vascular arcades to a point just below the macula (Fig. 5). This remained static for approximately 12 months, but the visual acuity decreased to 6/18 over this period.

Extensive screening investigations were carried out. Chest x-ray, liver scan, abdominal CT scan, and liver enzyme tests, failed to reveal any abnormalities. Systemic examination remained normal, and there was no evidence of hepatomegaly.

Because of the threat to vision in an only eye, it was decided to remove the tumour surgically despite little documented change in size of the mass itself. The excision was performed by Professor Wallace Foulds in Glasgow. His standard technique of a partial thickness scleral flap was used,¹³ the tumour being dissected off the retina. A sector iridectomy and anterior vitrectomy were performed.

Other than a small vitreous haemorrhage, which is not uncommon in these cases, the postoperative course was uneventful, the current visual acuity 3 months after surgery being 6/12.

Histology showed a malignant melanoma of mixed spindle B and epithelioid types (Fig. 6). The latter cells were atypical in that the cell boundaries were ill defined, and there was a marked variation in nuclear size and shape. Another interesting feature was the

Fig. 3 Right eye. Choroidal melanoma involving the pars plana.

Fig. 4 Right eye. Mid-phase fluorescein angiogram showing tumour vascularity.

presence of sheets of lymphocytes within the tumour mass.

Discussion

Studies in the United States have assessed the incidence of choroidal melanomas to be approxi-

Fig. 5 Right eye. Fundal painting showing the tumour inferiorly with serous retinal detachment and pigment deposition below the macula.

Fig. 6 Right eye. Choroidal melanoma of mixed spindle B and epithelioid types. Note infiltration by lymphocytes. ($\times 250$).

mately 5 to 7 new cases per million in the white population per year.¹⁴⁻¹⁶ Bilateral primary choroidal melanomas are even rarer, their incidence theoretically estimated at 0.55 cases per year in the United States.¹ One such bilateral case could thus be expected every 18 years in a population of 200 million whites. An explanation for the rare incidence of a choroidal melanoma developing in a second eye may well be the decreased life expectancy of the patients with choroidal melanoma, coupled with the fact that the incidence rate of choroidal melanomas increases with age, being highest between 60 and 70 years.¹ This does not allow much time for a second tumour to develop.

Other than the possibility of some systemic factor such as an immunological deficiency to account for bilateral tumours, it is important to exclude metastases from a primary skin melanoma, although melanoma metastatic to the eye has been reported in fewer than 40 cases.^{17,18} No evidence of a primary skin lesion was found in this case, and over the 4-year period of follow-up one might expect screening to detect additional sites of metastases if the lesion in the second eye was considered a secondary. Moreover, the histology of the tumours in the 2 eyes was different, the one being of spindle cell type and the other of mixed spindle B and epithelioid cells.

Evidence of a host immune response was present in the form of sheets of lymphocytes within the tumour mass in the second eye of this patient. This correlates well with the fact that the tumour had shown little tendency to enlarge while observed over a 2-year period. Recently Sergott and coworkers have shown that choroidal melanomata infiltrating lymphocytes have statistically significant different helper and suppression percentages as well as different functional characteristics from those of autologous peripheral blood lymphocytes and nontumour choroidal lymphocytes maintained in parallel cultures.¹⁹

This case of bilateral primary choroidal melanoma is the first to be reported in the United Kingdom and serves as a reminder of the importance of continued careful examination of the second eye for additional (and possibly treatable) pathology of any sort after the removal of one eye.

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References

- 1 Shamma HF, Watzke RC. Bilateral choroidal melanomas. Case report and incidence. *Arch Ophthalmol* 1977; **95**: 617–23.
- 2 Völcker HE, Naumann GDH. Multicentric primary malignant melanomas of the choroid: two separate malignant melanomas of the choroid and two uveal naevi in one eye. *Br J Ophthalmol* 1978; **62**: 408–13.
- 3 Rosen DA, Moulton GN. Multiple malignant melanomas in one eye. *Am J Ophthalmol* 1953; **36**: 73–5.
- 4 Sabates FN, Yamashita T. Congenital melanosis oculi complicated by two independent malignant melanomas of the choroid. *Arch Ophthalmol* 1967; **77**: 801–3.
- 5 Goldstein I, Wexler D. Melanosis uveae and melanoma of the iris in neurofibromatosis (Recklinghausen). *Arch Ophthalmol* 1930; **3**: 288–96.
- 6 Shine FW. Report of a case of bilateral sarcoma of the uveal tract. *NY Eye Ear Infirmary Clin Rep* 1930; **1**: 31–3.
- 7 Cordes FC, Cook RD. Simultaneous bilateral primary ocular malignant melanoma. *Trans Am Ophthalmol Soc* 1949; **47**: 80–92.
- 8 Wiesinger H, Phipps GW, Guerry D. Bilateral melanoma of the choroid associated with leukaemia and meningioma. *Arch Ophthalmol* 1959; **62**: 889–93.
- 9 Tade AA. Bilateral primary melanoblastoma of the choroid. *Vestn Ophthalmol* 1960; **4**: 30–5.
- 10 Buschmann W, Goder G. Das doppelseitige maligne Melanoblastom der Aderhaut. *Albrecht von Graefes Arch Klin Ophthalmol* 1964; **167**: 225–38.
- 11 Giarelli L, Silvestri F, Brandt M. Ein bilaterales Melanoblastom der Aderhaut, das beiderseits mit einem benignen Melanom vergesellschaftet ist. *Albrecht von Graefes Arch Klin Ophthalmol* 1972; **185**: 227–34.
- 12 Osborn EL, Walker JP, Weiter JJ. Bilateral choroidal melanoma: a case report. *Am J Ophthalmol* 1980; **12**: 1154–5.
- 13 Foulds WS. Experience of local excision of choroidal melanomata. *Trans Ophthalmol Soc UK* 1977; **97**: 412–5.
- 14 Cutler SJ, Young JL. *Third National Cancer Survey: incidence data*. National Cancer Institute Monograph no. 41. United States Government Printing Office, 1975.
- 15 Dorn HF, Cutler SJ. *Morbidity from cancer in the United States*. Public Health monograph no. 56. United States Government Printing Office, 1959.
- 16 Ganley JP, Comstock GW. Benign nevi and malignant melanomas of the choroid. *Am J Ophthalmol* 1973; **76**: 19–25.
- 17 Char DH, Schwartz A, Miller TR, Abele JS. Ocular metastases from systemic melanoma. *Am J Ophthalmol* 1980; **90**: 702–7.
- 18 Font RL, Naumann G, Zimmerman LE. Primary malignant melanoma of the skin metastatic to the eye and orbit. *Am J Ophthalmol* 1967; **63**: 738–54.
- 19 Sergott RC, McFall RC, Augsberger JJ, Shields JA, Folberg R, Nagy R. Growth and characterisation of choroidal melanoma infiltrating lymphocytes. *ARVO Abstracts* 1983; no 48: 191.