6. Albuquerque FC, Giannotta SL. Arachnoid cyst rupture producing subdural hygroma and intracranial hypertension: case reports. Neurosurgery 1997;41:951–6.
7. Gwan Go K, Houthoff H-J, Blaauw EH, Havinga P, Hartsuiker J. Arachnoid cysts of the Sylvian fissure. J Neurosurg 1984;60:803–13.

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Sir,

Massive extraocular extension as the presenting feature of a choroidal melanoma

It is uncommon for patients to present with extensive extraocular involvement as the primary presenting feature of uveal melanoma. However, in patients who are asymptomatic and do not seek ophthalmic evaluation or in those who refuse recommended treatment and allow tumour growth to continue, advanced progressive disease may occur. We report a case of a small choroidal melanoma presenting with massive extraocular extension, which was associated with minimal symptoms. Its appearance mimicked that of a lacrimal gland tumour. Despite the unusual clinical presentation, a diagnosis of melanoma was reached by biopsy of this extrascleral lesion.

Case report

A 58-year-old woman was referred to the Ophthalmic Oncology service at the Tennent Institute in Glasgow in October 1998. She had noticed a progressive enlarging mass on the superotemporal aspect of the left globe over the previous 3 months, which caused some discomfort. Past medical and ocular history were unremarkable. Ophthalmic examination revealed a visual acuity of 6/60 in the affected eye and a relative afferent pupillary defect. There was a pigmented bulbar mass in the superotemporal quadrant of the left globe restricting ocular movements (Fig. 1). Fundoscopy showed a small choroidal melanoma, 3 mm in height and 5 mm in diameter, in the posterior pole of the same eye. Examination of the right eye was unremarkable.

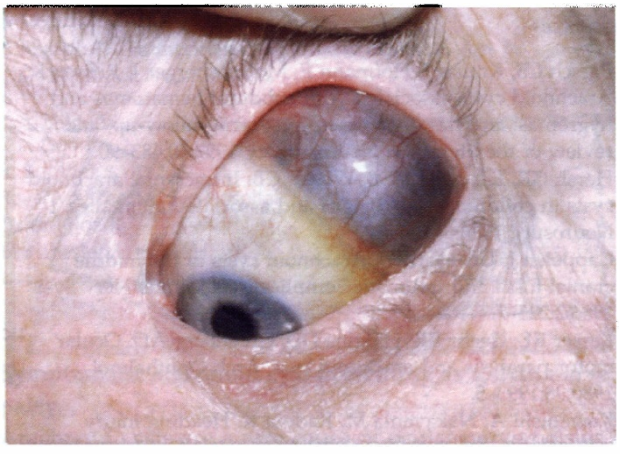

Fig. 1. *Pigmented bulbar mass in the superotemporal quadrant of the left globe at presentation.*

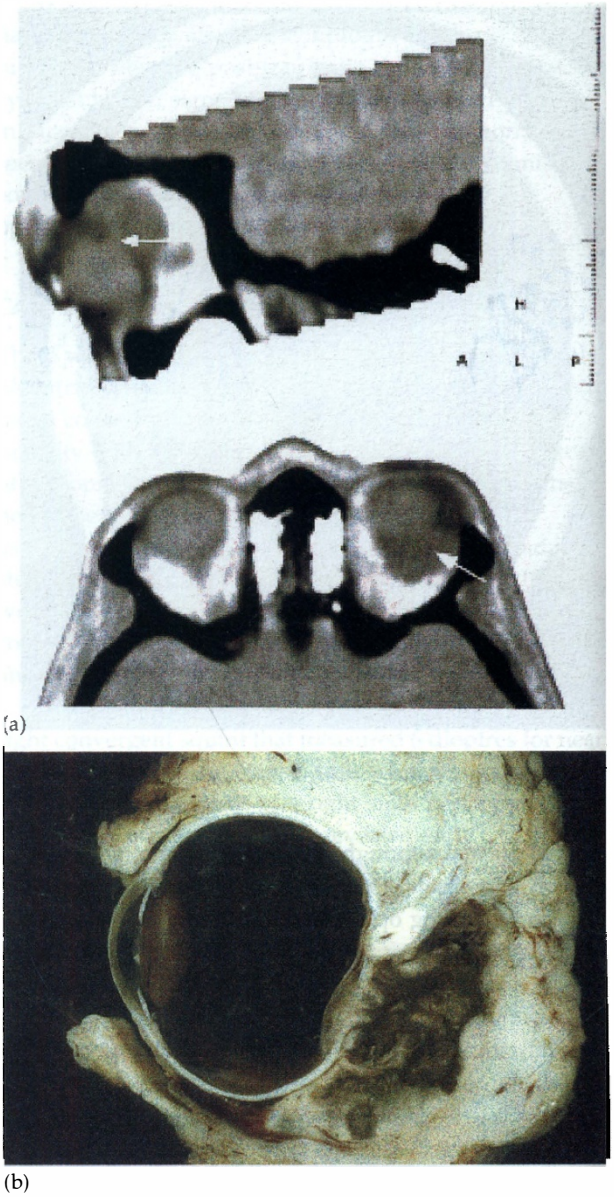

Fig. 2. *(a) CT scan demonstrating a defect in the sclera allowing continuity between intra- and extraocular contents (arrow). (b) Exenteration specimen showing massive extraocular spread..*

A CT scan confirmed a solid extraocular mass lying on the superolateral aspect of the left globe and extending back as far as the optic nerve. Differential diagnoses included extrascleral extension of a choroidal melanoma, a lacrimal gland tumour, a lymphoma, a pseudotumour or a metastatic deposit. The scan also demonstrated a defect in the sclera allowing continuity between intra- and extraocular contents (Fig. 2a). A metastatic screen including chest radiograph, liver function tests and liver ultrasound were normal.

The patient had an exenteration of the left orbit with preservation of eyelid skin. A peroperative biopsy of the extraocular mass revealed a spindle cell melanoma. Pathological examination of the exenteration specimen

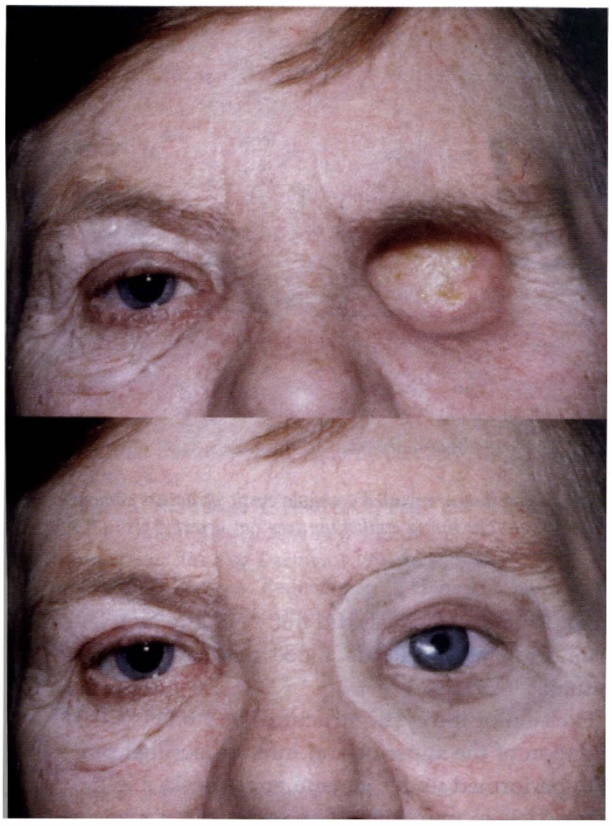

Fig. 3. *The healed left socket after exenteration (above) and after fitting of an adhesive retained prosthesis (below).*

confirmed a spindle cell choroidal melanoma with massive extraocular spread (Fig. 2b). Few mitotic figures were noted. The orbital tumour was widely cleared in the planes examined.

The patient made an uneventful recovery. The socket healed well over the following months. She was fitted with an adhesive retained prosthesis in the short-term (Fig. 3). She is currently awaiting titanium pegs to facilitate fitting of a Branemark osseointegrated prosthesis. Follow-up is 26 months and there is no evidence of local recurrence or distant metastases to date.

Comment

The reported incidence of extrascleral extension of uveal melanomas ranges from 10% to 15%.[1–3] These patients have a guarded prognosis regardless of the therapy chosen. Mortality with extrascleral extension has consistently been found to be 2–3 times greater than that for cases in which the tumour was confined totally within the eye at the time of enucleation.[1–3] Mortality from metastatic disease is shown to range from 66% to 85%.[1,2,4] Mortality is also increased when the tumour margin is surgically transected or non-encapsulated.[1–3] Large intraocular tumours and tumours of mixed or epithelioid cell type are more likely to be accompanied by extrascleral extension of the primary.[1] Our case does not obey these features in so far as the intraocular tumour was small and spindle in cell type.

Presenting symptoms and signs include a unilateral orbital mass, painless proptosis, decreased visual acuity, acute glaucoma crisis and intermittent diplopia.[5,6] Of interest in the present case are the relatively minor and non-specific symptoms experienced by the patient despite advanced disease.

The management of these patients is controversial, some proposing orbital exenteration and others believing that it does not increase survival. Those who advocate it in cases of extrascleral extension believe it to be effective in removing tumour which has not yet disseminated systemically. Those who oppose it feel that tumour extends into the orbit and disseminates via the systemic circulation concurrently, so that metastases are a *fait accompli* when extrascleral extension is identified. Shammas and Blodi[1] found that early exenteration for extrascleral extension was curative, with 87% of their patients surviving 5 years or more. However, Kersten *et al.*[7] reported that with longer follow-up of these patients and additional cases this form of treatment was palliative and prognosis was not altered in those who had an exenteration. Mean survival after exenteration for massive extraocular involvement in a recent report was 16 months.[6] Exenteration is effective, however, for local control of the disease and the associated intractable pain even if it does not improve patient survival.

References

1. Shammas HF, Blodi FC. Orbital extension of choroidal and ciliary body melanomas. Arch Ophthalmol 1977;95:2002–5.
2. Starr HJ, Zimmerman LE. Extrascleral extension and orbital recurrence of malignant melanomas of the choroid and ciliary body. Int Ophthalmol Clin 1962;2:369–85.
3. Affeldt JC, Minckler DS, Azen SP, *et al.* Prognosis in uveal melanoma with extrascleral extension. Arch Ophthalmol 1980;98:1975–9.
4. Wilder HC, Paul EV. Malignant melanoma of the choroid and ciliary body: a study of 2535 cases. Milit Surg 1951;109:370–8.
5. Rini FJ, Jakobiec FA, Hornblass A, *et al.* The treatment of advanced choroidal melanoma with massive orbital extension. Am J Ophthalmol 1987;104:634–40.
6. Lariakos S, Rapidis AD, Roumeliotis A, *et al.* Secondary orbital melanomas: analysis of fifteen cases. J Craniomaxillofac Surg 2000;28:148–52.
7. Kersten RC, Tse DT, Anderson RL, Blodi FC. The role of orbital exenteration in choroidal melanoma with extrascleral extension. Ophthalmology 1985;92:436–43.

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