

Cavernous haemangioma presenting as an orbital mass after enucleation for a choroidal melanoma: case report

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SUMMARY A patient presented with an ipsilateral orbital mass four years after enucleation for a choroidal malignant melanoma. Clinical examination revealed a darkly coloured mass suggestive of recurrent melanoma. CT scanning indicated a locally extensive tumour. Management was by excision without resort to orbital exenteration. Pathological examination revealed a cavernous haemangioma. The natural history of this tumour is discussed.

Cavernous haemangioma is the commonest primary orbital tumour.¹ Its pathogenesis is disputed, but it has been suggested that the neoplasm may become apparent only when growth takes place of a small, long-standing, or congenital pre-existing haemangioma.²

A case is reported in which a cavernous haemangioma arose in the orbit following enucleation of the ipsilateral eye for a uveal malignant melanoma. Intraoperative inspection of the socket during enucleation had revealed no evidence of an orbital tumour or vascular malformation.

Case report

A 63-year-old woman presented with sudden onset of blurred vision in the right eye. A thyroidectomy had been performed over 30 years previously for thyrotoxicosis, and there remained minimal lid retraction and lid lag on the right side, with slight proptosis. More recently she had developed Addison's disease, and had been treated with steroids. On examination the visual acuity in the right eye was 6/18 and in the left 6/9. In the right fundus there was a raised, lightly pigmented mass in the upper temporal quadrant above the macula, with an overlying serous retinal detachment. Systemic investigations revealed no evidence of a primary tumour elsewhere, and a diagnosis of amelanotic melanoma was made for this ocular tumour. Specific investigations for metastatic melanoma were negative.

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Growth of the tumour was recorded, and the visual acuity fell to 6/36 as a result of further accumulation of subretinal fluid. A right enucleation was therefore performed. Histological examination revealed a very lightly pigmented tumour situated in a postequatorial position in the temporal region and involving the macula. Microscopic examination disclosed a small, extremely lightly pigmented, spindle-cell, choroidal malignant melanoma. The associated retinal detachment was demonstrated pathologically. There was no evidence of extrascleral extension or involvement of

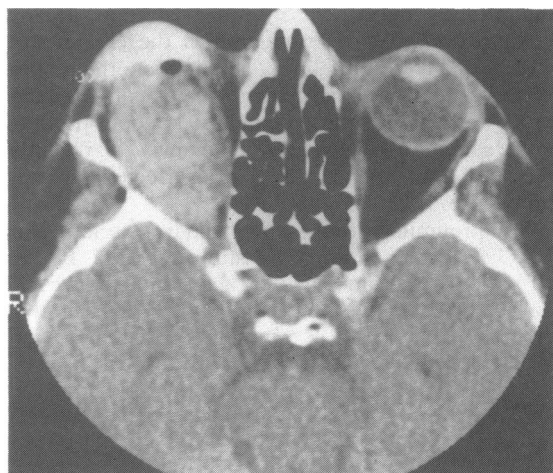


Fig. 1 CT scan showing large recurrent soft tissue mass lying behind the prosthesis.

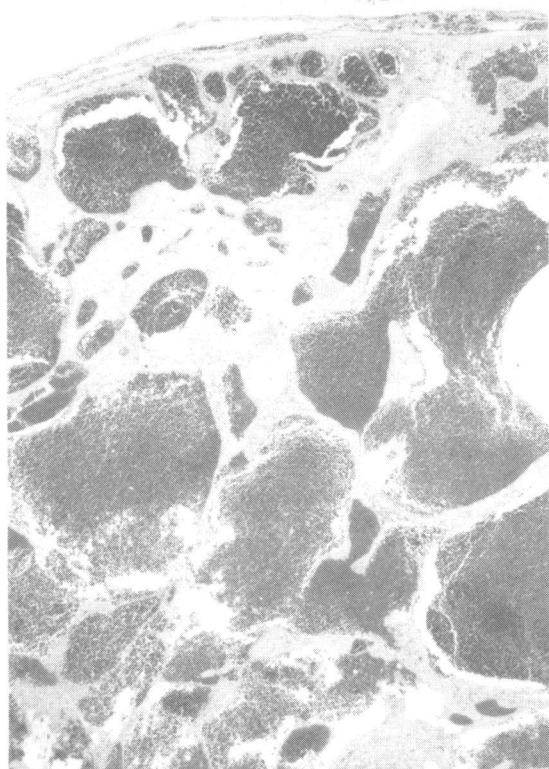


Fig. 2 Section of orbital mass showing numerous cavernous vessels filled with blood within a thin fibrous capsule typical of a cavernous haemangioma. Haematoxylin and eosin, $\times 36$.

the overlying retina. Subsequently a prosthesis was fitted.

Four years later the patient reported progressive forward displacement of the prosthesis. The clinical appearance was suggestive of a massive recurrence of melanoma in the enucleated right socket. A firm, bluish black mass was seen filling the superolateral aspect of the orbit. Staging investigations including full blood count, liver function tests (including gamma GT), chest x-ray, liver scan, and bone scan did not show any evidence of metastatic disease. A CT head scan delineated a large mass of soft tissue density with the appearance of recurrent melanoma. (Fig. 1). There was no evidence on the scan of any extension out of the orbital cavity.

The tumour appeared well circumscribed, and it was decided to attempt excision biopsy without resorting initially to orbital exenteration. Histological examination showed an encapsulated fibrous lesion within which there were numerous thin walled, endothelium lined, blood filled spaces. The appearances were typical of a cavernous haemangioma (Fig.

2). The histology of the enucleated eye was reviewed and confirmed a spindle B cell melanoma with no special features.

Discussion

Typically, cavernous haemangioma of the orbit arises in middle life,^{3,5} and it is commoner in women than in men.⁶ Usually the tumour presents with a long history of moderate, unilateral, painless protrusion of the eye, though occasionally proptosis may be marked or rapidly progressive.⁵ An orbital CT scan is the single most useful diagnostic test and shows a well circumscribed lesion with no osseous involvement.^{5,7}

Histologically cavernous haemangioma is quite different from capillary haemangioma of the orbit,⁸ and it is not considered likely that it arises by cavernous transformation of a capillary tumour present undetected from infancy.² Unlike capillary haemangiomas, orbital cavernous haemangiomas are discrete, well encapsulated tumours without large vascular tributaries.⁹ Large and small thin walled cavernous vessels identical to those within an orbital cavernous haemangioma have been found lying outside the tumour capsule. Apparent incorporation of some of these vessels within the tumour by newly formed reactive fibrous tissue has been observed, and it has been suggested that cavernous haemangiomas arise in the orbit from enlargement of previously existing abnormal vascular channels.⁹ The stimulus to this event may be disturbance in the blood flow within these vessels resulting from some extraneous event.⁹

The relevance of the present case is that no macroscopic evidence of abnormal vascular tissue within the orbit was detected during routine intra-operative inspection of the socket for evidence of extrascleral extension of uveal malignant melanoma, though a small pre-existing lesion may have been obscured by the orbital fat. The haemangioma described expanded rapidly almost to fill the orbit within four years, and this behaviour would seem to exclude steady growth of a congenital tumour as the natural history of this particular lesion. Atypical, rapid growth of cavernous haemangioma does occur but is uncommon.⁵ It is tempting to speculate whether the absence of the globe in the unusual case reported here permitted rapid expansion of the tumour.

The importance of not assuming local recurrence of malignant melanoma but of obtaining further tissue for histological examination is emphasised. In this case it avoided unnecessarily radical treatment by orbital exenteration or inappropriate radiotherapy.

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