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Benign fibrous histiocytoma of the choroid

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Abstract

Benign fibrous histiocytoma (BFH) is a common soft tissue tumour. However, its occurrence in the orbit was not well recognised in the past. It is only in recent years that this condition has become more frequently diagnosed. Now, it is believed to be the commonest mesenchymal tumour of the orbit in adults. Its occurrence in the uveal tract is, however, exceedingly rare. Only one case of presumed fibrous histiocytoma of the choroid has been reported in the literature. We report herein a case of BFH of the choroid in the left eye of a Chinese woman. The patient presented with a huge but asymptomatic raised choroidal mass. Results of choroidal biopsy showed no sign of malignancy but definitive diagnosis could not be made. Enucleation was finally performed. The diagnosis was made on detailed evaluation of the results of the immunohistochemical staining and the ultrastructural findings. The patient remained well at the latest follow-up, which was 33 months after enucleation. Although BFH of the choroid is very rare, its benign nature and the availability of choroidal biopsy for tissue diagnosis make it important to include this as one of the differential diagnoses for amelanotic choroidal mass.

Key words Benign, Fibrous, Histiocytoma, Choroid, Amelanotic

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Benign fibrous histiocytoma is a tumour of histiocytes and fibroblasts. It is commonly found in soft tissues[1] and is believed to be the commonest mesenchymal tumour of the orbit in adults.[2,3] However, its occurrence in the uveal tract is exceedingly rare. We report herein a case of benign fibrous histiocytoma of the choroid, which to our knowledge is the second case reported in literature.

Case report

A 69-year-old Chinese woman presented to the Eye Department at the Prince of Wales Hospital in 1994 with a 1-year history of gradual deterioration of vision of the left eye. There were no other associated ocular or systemic symptoms. Her past ocular and medical history were unremarkable. Examination revealed a visual acuity of 20/70 and 10/200 in the right and left eyes respectively. Apart from moderate cataract in both eyes, there was a large solitary yellowish-white raised choroidal lesion measuring about 6 disc diameters and extending from mid- to peripheral retina in the infero-temporal quadrant. This mass did not transilluminate and there was no accumulation of lipofuscin pigment, no exudative retinal detachment or choroidal folds. The rest of the ocular examination was also negative.

B-scan ultrasonography confirmed the grossly elevated choroidal lesion affecting mainly the temporal side of the inferior half of the fundus (Fig. 1). There was no obvious choroidal excavation or acoustic hollowness. A standardised A-scan to assess the acoustic structure was not available. Fluorescein angiography showed irregular filling of the mass in the arterial and arteriovenous phases with hyperfluorescence over areas of atrophic retinal pigment epithelium. There was diffuse and prolonged pooling of dye within the mass at the later phases (Fig. 2). No double circulation was noted. A systemic investigation, including chest radiograph, complete blood picture, ESR, liver and renal function tests, was negative. Computed tomography (CT) showed confinement of the tumour within the left globe and no calcification was noted.

As the clinical picture was compatible with a localised amelanotic melanoma, choroidal biopsy (incisional method) under local anaesthesia was performed. Pathological examination of the specimen could not confirm the presence of mitosis but cells were slightly hyperchromatic. The cells had very little cytoplasm and looked mesenchymal in origin. No melanin could be seen. The S100, demsin, CAM 5.2, AE1/AE3, leucocytic common antigen and neurone-specific enolase stains were all negative (Table 1). Although the negative S100 finding made melanoma and other benign neural tumours unlikely, a definitive histological conclusion could not be reached. The patient was put under close clinical observation. With progression of the cataract and further drop in vision to 3/60

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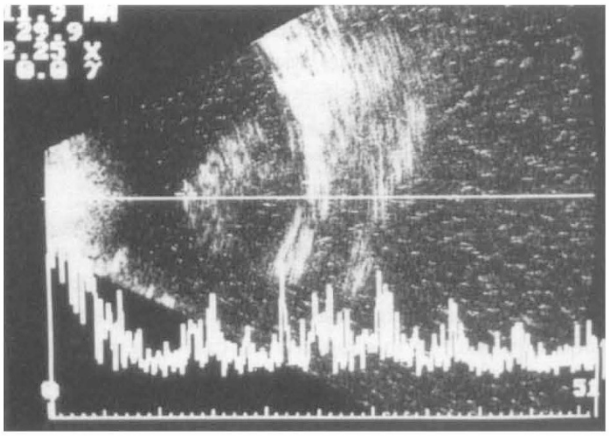

Fig. 1. *Ultrasonography (B-scan) demonstrating the large raised choroidal mass.*

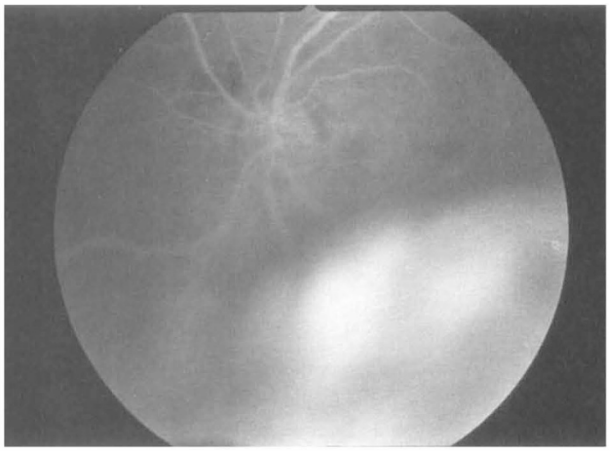

Fig. 2. *Pooling of fluorescein dye within the mass at the late venous phase of fluorescein angiography.*

3 months later, the patient chose enucleation instead of clinical observation to obviate the risks of an ocular neoplasm. She made an uneventful recovery and has remained well up to the time of this report, 33 months after the operation.

Pathological evaluation

The enucleated eye measured 2.5 cm in diameter. A well-circumscribed whitish choroidal tumour ($10 \times 11 \times 15$ mm) was found in the infero-temporal quadrant. It was attached and confined to the inner surface of the sclera. The tumour was covered by retinal tissue. Cut section showed homogeneous white firm tissue. Necrosis was not seen on gross examination.

Histologically, the tumour displayed a uniform appearance. It consisted of interlacing fascicles of spindle cells supported in a dense collagenous stroma (Fig. 3). Focal hyalinisation was seen in the centre. The vascularity of the tumour was low. The spindle cells had bland oval nuclei, indistinct nucleoli and a scanty amount of eosinophilic cytoplasm. Mitosis and necrosis were absent. There was no invasion into sclera. The optic nerve was spared. The retinal pigment epithelium overlying the tumour was stretched but otherwise unremarkable.

Immunohistochemical staining was performed on the paraffin sections, employing a panel of antibodies (Table 1), using the microwave antigen retrieval method and avidin–biotin–peroxidase technique, with appropriate positive and negative controls. The tumour cells showed intense positive staining for vimentin, and negative staining for S100 protein, HMB-45 (markers for melanoma), muscle-specific actin, desmin (markers for smooth muscle tumour), CAM 5.1, AE1/AE3 (markers for epithelial tumour), KP1, MAC 387 and HAM 56 (markers for histiocytes). The genuine negative staining of the tumour cells for these antibodies was supported by the positive internal controls in the sections, namely positive staining of the nerves for S100 protein, positive staining of the smooth muscle in the wall of blood vessels for muscle-specific actin and desmin, and positive staining of the retinal pigment epithelium for CAM 5.2 and AE1/AE3.

Ultrastructural examination revealed that the spindle tumour cells showed features of fibroblastic differentiation, containing abundant rough endoplasmic reticulum; they were supported in a stroma rich in collagen fibres (Fig. 4). In addition, there were occasional cells that contained a number of lysosomal granules, consistent with histiocytic differentiation.

The diagnosis of a benign fibrous histiocytoma, arising from the choroid of the eye, was thus made.

Table 1. *Antibodies employed in the present study and their staining reactions*

Antibody	M/P	Source	Dilution	Staining reaction
Vimentin	M	Dakopatts, Copenhagen, Denmark	1:100	Positive
S100 protein	P	Dakopatts, Copenhagen, Denmark	1:100	Negative
HMB-45	M	Enzo Biochem, New York, NY	Undiluted	Negative
Muscle-specific active (HHF) 35)	M	Enzo Biochem, New York, NY	Undiluted	Negative
Desmin	M	Dakopatts, Copenhagen, Denmark	1:100	Negative
CAM 5.2	M	Becton Dickinson, San Jose, CA	1:5	Negative
AE1/AE3	M	Hybritech, San Diego,CA	1:400	Negative
KP1	M	Dakopatts, Dako Diagnostic, Hamburg, Germany	1:2000	Negative
MAC 387	M	Dakopatts, Dako Diagnostic, Hamburg, Germany	1:100	Negative
HAM 56	M	Dakopatts, Dako Diagnostic, Hamburg, Germany	1:50	Negative

M, monoclonal; P, polyclonal.

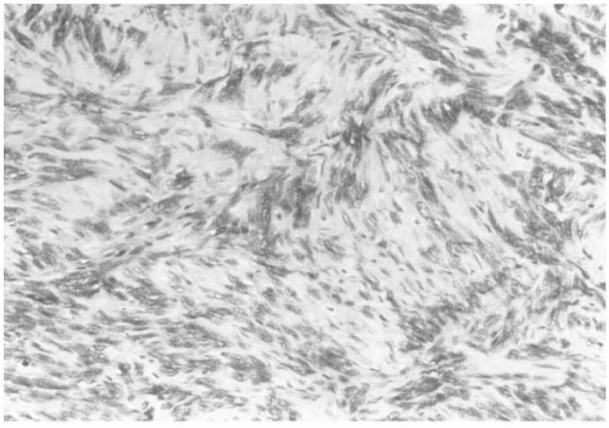

Fig. 3. *The tumour shows a uniform histological appearance, consisting of interlacing fascicles of spindle cells supported in a dense collagenous stroma (haematoxylin & eosin, ×150).*

Discussion

Fibrous histiocytoma is a benign soft tissue tumour composed predominantly of fibroblastic cells arranged in a storiform pattern or interlacing fascicles, supported in a collagenous stroma and admixed with varying numbers of histiocytes, foam cells, siderophages and inflammatory cells. Immunohistochemically, the proliferating spindle fibroblastic cells stain positively for vimentin but usually negative for lysozyme and other histiocytic markers including MAC 387.[4] This tumour most commonly occurs in the dermis and superficial subcutis, but is also found in deep soft tissue and occasionally in parenchymal organs.

In contrast to its relatively common occurrence in the orbit, documented cases of benign fibrous histiocytoma within the globe are very rare. To our knowledge, only one such case has been reported in the English literature.[5] That case of presumed fibrous histiocytoma of the choroid affected a 32-year-old woman, whose clinical diagnosis before enucleation was a solitary choroidal haemangioma. She eventually developed total secondary retinal detachment and neovascular glaucoma that necessitated enucleation. The histological,

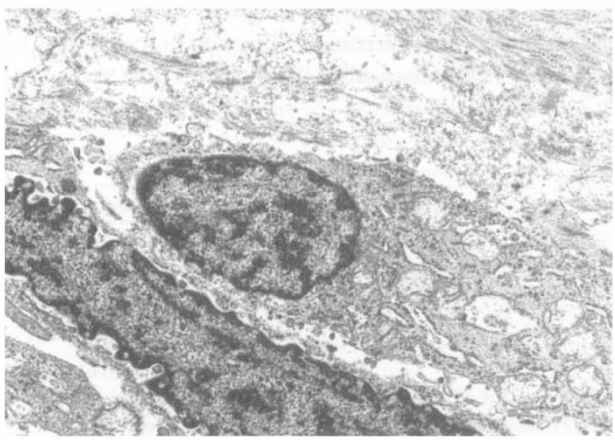

Fig. 4. *Ultrastructurally, the tumour cells contain abundant rough endoplasmic reticulum, and the stroma is rich in collagen fibres (×6000).*

immunohistochemical and ultrastructural features of that tumour are similar to ours apart from the presence of a plaque of osseous metaplasia on the surface of the tumour that was associated with a pseudotubular arrangement of overlying hyperplastic pigment epithelium.

As osseous metaplasia is not uncommon in the superficial region of circumscribed choroidal haemangiomas, there remains a distinct possibility that the reported case might have been a sclerosed choroidal haemangioma.[6] In this regard, our case was unique in that it exhibited a uniform histological appearance, and the immunohistochemical and ultrastructural findings supported its being a genuine case of primary benign choroidal fibrous histiocytoma.

Differential diagnoses of benign choroidal fibrous histiocytoma include other spindle cell tumours described in that location, notably amelanotic melanoma, leiomyoma, neurofibroma and neurilemmoma.

Spindle cell melanoma commonly occurs in the anterior uveal tract. The tumour cells of the reported cases in the posterior uveal tract usually showed more prominent pleomorphism and large nucleoli. Moreover, they stained positively for S100 protein and HMB-45, allowing their differentiation from benign fibrous histiocytoma. Leiomyoma, a benign smooth muscle neoplasm, consists of spindle cells with cigar-shaped nuclei and eosinophilic fibrillary cytoplasm that shows positive immunostaining for actin and desmin. Peripheral nerve sheath tumours, namely neurofibroma and neurilemmoma, possess distinctive histological appearances. The former comprises wavy spindle cells embedded in a myxoid stroma, while characteristic Antoni type A and B patterns, together with hyalinised vessels, constitute the histological hallmark of the latter. Furthermore, peripheral nerve sheath tumours show positive immunostaining for S100 protein.

In clinical practice, we have to consider other differential diagnoses such as choroidal haemangioma, metastatic carcinoma, lymphoma, granuloma, and giant posterior scleritis.[7] Choroidal biopsy[8,9] may be required when the diagnosis cannot be made with certainty by systemic investigation and non-invasive ancillary diagnostic procedures such as A- and B-scan ultrasonography, fluorescein angiography, computed tomography and magnetic resonance imaging. The risk of enhanced systemic and local spread of the neoplasm is a deterrent to performing a choroidal biopsy. However, Foulds *et al.*[10] have demonstrated that the added risk of either systemic or local spread is low. The biopsy can be achieved with fine needle aspiration or wedge biopsy technique. We chose the wedge biopsy approach as the mass was rather peripherally located and easily accessible without detaching any extraocular muscles.

In accordance with its counterpart in the soft tissue of other parts of the body, we believe the behaviour of fibrous histiocytoma of the choroid is benign and it may be feasible for early cases to be managed with local resection or radiotherapy.[11] Retrospectively, the biopsy results of our patient were compatible with the diagnosis

of benign choroidal fibrous histiocytoma. However, this was such a rare entity that it had not been considered until after enucleation. We recommend the addition of this tumour to the list of differential diagnoses for amelanotic choroidal mass.

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References

1. Enzinger FM, Weiss SW. Soft tissue tumours. 1st ed. St Louis: CV Mosby, 1988:284–300.
2. Jakobiec FA, Font RL. Orbit. In: Spencer WB, editor. Ophthalmic pathology: an atlas and textbook. 3rd ed. Philadelphia: Saunders, 1986:2568–80.
3. Shields JA. Fibrous connective tissue tumours. In: Shields JA. Diagnosis and management of orbital tumours. 1st ed. Philadelphia: Saunders, 1989:199–210.
4. Fletcher CD. Benign fibrous histiocytoma of subcutaneous and deep soft tissue: a clinicopathological analysis of 21 cases. Am J Surg Pathol 1990;14:801–9.
5. Croxatto JO, D'Allessandro CD, Lombardi A. Benign fibrous tumour of the choroid. Arch Ophthalmol 1989;107:1793–6.
6. Shields JA, Shields CL. Vascular tumours of the uvea. In: Shields JA, Shields CL, editors. Intraocular tumours: a text and atlas. Philadelphia: Saunders, 1992:240–52.
7. Benson WE, Shields JA, Tasman W, Crandall AS. Posterior scleritis: a cause of diagnostic confusion. Arch Ophthalmol 1979;97:1482–6.
8. Scherfig E, Prause JU, Jensen OA. Transvitreal retinochoroidal biopsy. Graefes Arch Clin Exp Ophthalmol 1989;227:369–73.
9. Fastenberg DM, Finger PT, Chess Q, Koizumi JH, Packer S. Vitrectomy retinotomy aspiration biopsy of choroidal tumours. Am J Ophthalmol 1990;110:361–5.
10. Foulds W, Lee W, Roxburgh D, Damato BE. Can chorio-retinal biopsy be justified? Trans Ophthalmol Soc UK 1985;104:864–8.
11. Shields JA. Fibrous connective tissue tumours. In: Shields JA, editor. Diagnosis and management of orbital tumours. Philadelphia: Saunders, 1989:300–3.