

Choroidal osteoma presenting in pregnancy

B K McLEOD

From the West of England Eye Infirmary, Exeter

SUMMARY A case of choroidal osteoma presenting during pregnancy is reported. The clinical findings are summarised and the features of choroidal osteoma are discussed.

A 32-year-old woman presented with a history of occasional blurring of vision of the left eye over the period of 10 weeks. These attacks had lasted for a few minutes and cleared, leaving her with no visual defect. During the month prior to presentation, however, she had noticed a persistent blurring of the central vision. There was no relevant medical history apart from the fact that she was in the third month of an uncomplicated pregnancy.

Her corrected visual acuities were 6/4 in the right eye and 6/6 in the left eye. The left visual field showed a relative scotoma extending from the blind spot towards fixation. Examination of the right eye showed it to be entirely normal, but the left eye had a flat, well circumscribed, yellowish, mottled, juxta-papillary lesion extending from the inferior aspect of the disc upwards to involve the macula (Fig. 1). Fine tufts of irregular vessels were seen on the surface of the lesion.

Correspondence to B K McLeod, FRCS, Department of Ophthalmology, Clinical Sciences Building, Leicester Royal Infirmary, PO Box 65, Leicester LE2 7LX.

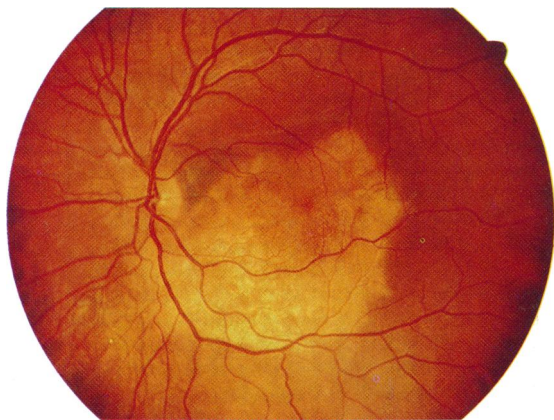


Fig. 1 Posterior pole of the left eye showing large juxtapapillary choroidal lesion.

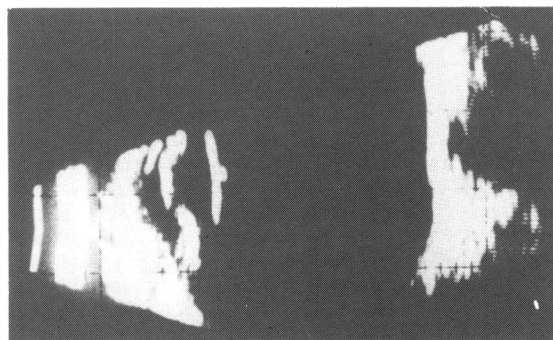


Fig. 2A Ultrasound B scan of the left eye showing the tumour with acoustic shadowing of the structures behind it.

Ultrasound B scan showed an acoustically dense choroidal mass at the posterior pole which caused an acoustic shadow. On reduction of the gain which suppressed all other posterior wall echoes, a strong echo persisted from the mass (Figs. 2A, B). The patient was kept under regular review, but no other investigations were performed until she had given birth at term. On re-examination a slight increase in the size of the lesion was noted, but the visual acuity and the scotoma remained static. A fluorescein angiogram showed early patchy hyperfluorescence of the lesion, which persisted into the later stages of the angiogram. A CT scan showed a radiodense lesion

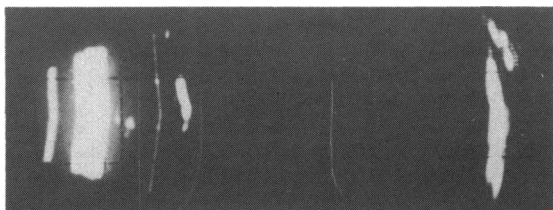


Fig. 2B Ultrasound B scan of the same eye showing a highly reflective tumour at low ultrasonic intensity.

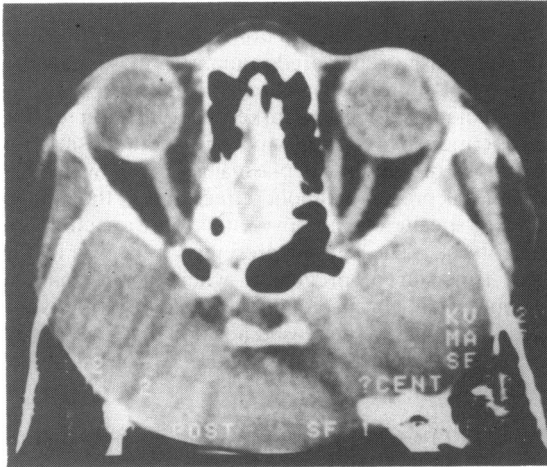


Fig. 3 Computerised axial tomography scan showing the presence of a bony plaque at the posterior pole of the left eye.

adjacent to the left optic nerve head, and this had the same density as normal bone (Fig. 3). A diagnosis of choroidal osteoma was made. The patient has been followed up for over a year, with no further visual deterioration or enlargement of the lesion being demonstrated.

Discussion

CLINICAL FEATURES

The features of choroidal osteoma were described by Gass and Guerry¹ when they described four young female patients with similar juxtapapillary lesions and sharing similar clinical characteristics. In a separate report on one of these patients Williams and Fort² reported the pathological findings on one of the eyes enucleated for a suspected choroidal melanoma after a positive ³²P test.

Choroidal osteoma tends to occur in young females patients. In a series of 15 cases reviewed by Gass³ 14 were female, with an age range of 8–36 and a median age of 20 years. Unilateral involvement is usual, with no preference for it to affect either eye having been found. However, bilateral involvement has been reported,³ and multiple occurrences in a patient with recurrent orbital pseudotumour are known.⁴

Blurred vision and metamorphopsia are the main presenting features, and paracentral scotoma may be demonstrated. The visual acuity is usually good unless extensive macular complications have occurred. The lesions have an affinity for the juxtapapillary region and tend to have irregular borders and a geographic pattern, with a yellow-white or orange-red colour, depending on the degree of thinning of the retinal pigment epithelium. Multiple short tufts

of branching vessels on the tumour surface arising from marrow spaces within the osteoma are said to be characteristic. The prompt perfusion of the chorio-capillaris gives rise to diffuse mottled hyperfluorescence early in the fluorescein angiogram, and later patchy fluorescein staining may be due to filling of these marrow spaces where they communicate with the surface.

The typical ultrasound finding is that of an acoustically dense, slightly elevated choroidal mass which renders the tissue behind it anechoic, producing an apparent defect in the retrolbulbar tissue. If the ultrasound gain is reduced, so that all the posterior wall echoes are suppressed, a high-amplitude echo persists from the osteoma. Orbital x-rays may show the radiodense lesion, which may be highlighted by polytomography. CT scan of the orbit will best show the lesion of the posterior pole of the eye with a density similar to that of bone.

NATURAL HISTORY

The visual acuity may be good, with minimal visual disturbance at first, but lesions near the macula may give rise to serous retinal detachment and subretinal neovascularisation. Of 14 eyes followed up for seven years Gass reported that eight maintained 6/12 vision while six fell to 6/60 or worse.³

HISTOPATHOLOGY

The features of two eyes studied after enucleation have been reported and another case reviewed, having been reported to be an ossified haemangioma by Reese.⁴ The lesions were situated between the inner one-third and the outer two-thirds of the choroid, separated from the retinal pigment epithelium and Bruch's membrane by a thin layer of fibrous tissue. The retinal pigment epithelium showed patchy thinning and depigmentation but no hyperplasia, and in these areas of disruption rod and cone loss was found, with a layer of subretinal exudate. No evidence of haemangioma or changes suggesting the origin of the osseous tissue was found.

ORIGIN

The origin of choroidal osteoma is still a cause for discussion. Are they primary lesions, or is there an underlying stimulus for ossification? It is true that choroidal haemangiomas undergo osseous metaplasia, but this always occurs on the inner surface of the haemangioma in association with extensive degenerative and proliferative changes of the retinal pigment epithelium not seen in the osteomas examined so far. Heterotopic bone formation is seen in degenerated eyes following trauma, inflammation, and long standing retinal detachment. However,

hyperplastic changes of the retinal pigment epithelium are usually seen, and the origin of the bone is thought to be due to metaplasia of the retinal pigment epithelium in response to degenerative changes. Choroidal haemorrhage and focal choroiditis, which might be implicated in bone formation, have not been reported in previous cases of choroidal osteoma and were certainly not a feature of the case reported in this paper. Katz and Gass⁵ reported a case of multiple choroidal osteoma occurring in a 10-year-old girl with recurrent orbital pseudotumour, and during the period of active growth of the choroidal lesions they recorded transient elevation of the serum parathyroid hormone level and alkaline phosphatase. But there are few other recordings of an underlying metabolic disorder, and routine biochemical studies have been found to be unhelpful.

The optic nerve head has a complicated embryological development and is a common site for developmental tumours such as melanocytomas, astrocytic hamartomas, and choroidal haemangiomas. However, choroidal osteomas show features atypical of developmental tumours, such as their exclusive location in the juxtapapillary choroid, their occurrence predominantly in females, and their capacity for growth in later life. Some authorities now believe that the structure of the osteomas and their ability to develop in previously unaffected eyes indicate that they develop from reactive metaplasia

of mesenchymal or retinal pigment epithelial cells. The stimulus for this reaction is not clear, but since the lesions occur predominantly in females a hormonal influence has been postulated.^{3,5} During pregnancy there is a marked modification of the endocrine environment and the metabolism—for example, the increase in plasma oestrogen, which has a general protein anabolic effect and is thought to have a specific stimulatory effect on the activity of osteoblasts. The patient presented here was in the first trimester of pregnancy when her permanent symptoms first occurred, and it would be attractive to postulate that the increased hormonal activity might be implicated in the exacerbation of the choroidal osteoma.

I thank Mr J Thomsitt for permission to report this case and Miss K Jones for the photography.

References

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