Complete infarction of the eye complicating a choroidal malignant melanoma

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Abstract

Infarction of malignant melanoma of the choroid has been previously reported, but infarction of the whole eye in association with infarction of melanoma is a rare event that has not been previously described. We present such a case and discuss the possible pathogenesis.

Choroidal malignant melanoma is the commonest intraocular tumour in adults, becoming more frequent with increasing age.¹ Such tumours are known to undergo partial or complete infarction, and various autoimmune mechanisms have been postulated.²⁻⁴ In the present case infarction of a choroidal melanoma was associated with infarction of most of the globe.

Case report

An 83 year old man presented to the casualty department of Southampton Eye Hospital complaining of blurred and distorted vision in the right eye for several weeks. Fundal examination revealed a large, inferotemporal pigmented mass extending to the optic disc with an associated exudative retinal detachment. Ultrasonography of the right eye confirmed a solid lesion, which was presumed to be primary choroidal malignant melanoma. The left fundus was normal. General examination, full blood count, erythrocyte sedimentation rate, liver function tests, and chest x ray revealed no abnormality.

He elected to be managed conservatively and was to be reviewed in three months. Two months later, however, he returned to the casualty department complaining of a three-day history of deteriorating vision in the right eye culminating in severe pain and complete loss of vision (no perception of light). The intraocular pressure was 70 mmHg, with corneal oedema and a fixed, dilated, oval pupil. A diagnosis of glaucoma secondary to choroidal malignant melanoma was made. The eye was enucleated four days later, by which time the anterior chamber had developed a total hyphaema. Postoperative recovery was uneventful. Three months' follow-up revealed no recurrent tumour.

PATHOLOGY

Macroscopic inspection of the eye sectioned parasagittally revealed a completely detached retina with retroretinal haemorrhage. Dense connective tissue was adherent to the outside of the eye posteriorly.

Histological preparations showed a pigment-containing malignant melanoma arising from the

Figure 1 A diagram of a parasagittal section of the eye showing the location of the tumour (T), the detached retina (R), and retroretinal haemorrhage (H). The florid fibroblastic reaction extends from the region of the optic nerve (F) deep to the insertion of inferior oblique (IO) and into the tendon of the inferior rectus (IR). Optic nerve (N), lens (L), ciliary body (C).

region adjacent to the optic disc and extending midway into the vitreous chamber. The retina was detached partly by tumour and partly by haemorrhage (Fig 1). Fresh blood filled the

Figure 2 Infarcted ciliary body (top) and sclera. The outer third of the sclera shows a polymorphonuclear leucocyte infiltrate (P). (Haematoxylin and eosin, $\times 60$.)

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Figure 3 Connective tissue around the optic nerve and ciliary arteries showing the prominent fibroblastic reaction. Inset: illustrates the bizarre nature of the cells and a mitotic figure (M). (Haematoxylin and eosin, $\times 85$, inset $\times 215$.)

anterior chamber, and the lens showed cataractous change. No anterior synechiae were seen, and no cell debris could be identified in the trabecular meshwork.

Except for the cornea, all intraocular structures were totally necrotic, and they could be identified only by ghost-like profiles of the original cells (Fig 2). The inner two-thirds of the sclera was necrotic and acellular, and a band of pyknotic polymorphonuclear leucocytes was seen in the outer third of the sclera (Fig 2). Somewhat bizarre stellate and elongated cells with abundant slightly basophilic cytoplasm and large hyperchromatic nuclei with occasional mitoses were seen in the extraocular adipose tissue round the optic nerve (Fig 3). Such cells extended as a thin layer deep to the scleral attachment of the inferior oblique muscle and were seen throughout the tendon of the inferior rectus muscle. Although separated by the intervening sclera, the bizarre cells were related to the choroidal attachment of the necrotic malignant melanoma. These bizarre cells could, however, be distinguished from malignant melanoma cells by the absence of S100 protein expression immunohistochemically and by the lack of melanosomes on electron microscopy. From their distribution round the eye and their histological characteristics the bizarre cells were identified as a particularly florid fibroblast reaction to the ocular infarction.

No evidence of vasculitis or intravascular thrombi was detected in vessels of the sclera, optic nerve, or periocular tissue.

Discussion

Although infarction of malignant melanomas of the choroid is well documented,² total infarction of the eye with a malignant melanoma has not, to the best of our knowledge, been described previously. The mechanism for infarction in this case is unclear. As multiple arteries supply the eye (Fig 4), infarction is not likely to be due to

Figure 4 Diagram of the arterial supply to the eye. Central retinal artery (C). Short (S) and long (L) posterior ciliary arteries pass circumferentially around the eye and could have been occluded by high intraocular pressure, thus causing infarction of intraocular structures in the present case. Anterior ciliary artery (A) and posterior conjunctival arteries (P).

arterial embolic occlusion. Lack of vascular congestion and massive haemorrhage argue against venous infarction. From the history it appears that vision failed seven days before the eye was removed. It seems probable that a combination of glaucoma and infarction of the malignant melanoma resulted in a massive rise in intraocular pressure (measured at 70 mmHg). At such a pressure it is possible that the choroidal arteries on the inner aspect of the sclera (Fig 4) were compressed, resulting in infarction of virtually all the intraocular structures.

The major pathological feature, apart from the necrosis, was the florid fibroblast proliferation in connective tissue round the optic nerve and attachments of the inferior oblique and inferior rectus muscles. Despite the bizarre nature of some of the fibroblasts they could be distinguished from metastatic melanoma cells by immunohistochemistry and electron microscopy. The fibroblast reaction suggests that infarction of the eye occurred five to seven days before enucleation, the time when vision started to fail.

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