Coagulative necrosis in a malignant melanoma of the choroid at the macula with extensive subretinal hemorrhage

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The authors present a case report and pathologic examination of a malignant melanoma of the choroid located at the macula, with the unusual development of extensive subretinal hemorrhage which obscured the clinical diagnosis. The cause of the subretinal hemorrhage was demonstrated to be coagulative necrosis of the tumor produced by invasion and occlusion of a choroidal artery by neoplastic cells.

The clinical diagnosis of malignant melanoma of the choroid is often difficult. In a pathologic study by Ferry of 529 eyes with clear media, which were enucleated following a diagnosis of malignant melanoma of the choroid, 19 per cent of the eyes did not contain melanomas. The differential diagnosis of small dark lesions at the macula is extremely difficult because the ophthalmoscopic appearance of malignant melanoma and of hemorrhagic disciform macular degeneration can be similar. Subretinal hemorrhages in or around small macular lesions, however, are characteristic of disciform macular degeneration, and most authors believe they are rare in malignant melanoma. This report illustrates a case in which the development of extensive subretinal hemorrhage adjacent to a small macular mass indicated the diagnosis of disciform macular degeneration, but pathologic examination of the enucleated eye demonstrated an infarcted malignant melanoma of the choroid. In addition, pathologic examination established the cause of the infarction and the subretinal hemorrhage.

Case report

A 78-year-old man was examined at the Jules Stein Eye Institute in December, 1971, one month after he first noted blurring of central vision of his right eye. The visual acuity of the right eye was 20/100, and a slightly elevated, solid dark subretinal mass was found at the macula. The surface of the mass was coarsely mottled with clumps of orange pigment. No subretinal hemo-
of the posterior portion of the globe showed from the patient’s history or physical examination and laboratory tests revealed no evidence of metastases or other extracocular neoplasms.

During the next 11 months, the visual acuity of the right eye deteriorated to 2 ft./200, and the subretinal mass enlarged toward the optic disc. A small exudative retinal detachment was present inferiorly near the ora serrata. Two days after an examination in November, 1972, the patient noted a sudden deterioration of vision in the superior visual field of the right eye. Examination four days later revealed an extensive bullous retinal detachment extending from the inferior margin of the subretinal mass to the ora serrata inferiorly. The subretinal fluid was dark grey and obscured the underlying choroid (Fig. 1). Dark-red subretinal hemorrhage was present superior to the mass. There was no hemorrhage or inflammation in the vitreous.

Although the sudden occurrence of extensive subretinal hemorrhage made the diagnosis of malignant melanoma of the choroid less likely, the patient requested removal of the eye, since the presence of a malignant tumor could not be completely ruled out. A repetition of thorough general physical examination and laboratory tests revealed no evidence of extracocular neoplasms. Nothing from the patient’s history or physical examination suggested a bleeding diathesis, and results of laboratory tests, including platelet count, partial thromboplastin time, and prothrombin time, were normal. During enucleation of the right eye, no evidence of extracocular tumor was found.

Pathologic examination. External examination revealed a right eye of normal shape measuring 24.7 mm. in anteroposterior diameter, with 4.5 mm. of attached optic nerve. Transillumination of the posterior portion of the globe showed no light transmission in the temporal and inferior sectors. The episclera in these sectors was unremarkable. The eye was sectioned anteroposteriorly above the optic nerve through the 11 and 4 o’clock meridians.

Stereomicroscopic examination showed a dark choroidal mass, measuring 7.8 mm. (horizontal) by 9 mm. (vertical), in the region of and superior to the macula (Fig. 2). On the cut surface the tumor was biconvex in shape with a maximum thickness of 1.5 mm.; a circular area, centrally, was lighter in color than the surrounding brown mass. The subretinal space overlying the mass and extending throughout the inferior sectors to the ora serrata contained clotted and degenerating blood (Fig. 3). The detached retina overlying the mass and the subretinal hemorrhage was rumpled but intact. Multiple blocks through the base of the tumor were embedded in paraffin and microsectioned.

Microscopic examination revealed a circumscribed melanoma composed entirely of epithelioid cells in a nodular pattern (Fig. 4). In a few areas outlying the mass, tumor cells were found in small scleral canals, but the episclera was not involved. Sections taken near the cut surface showed central coagulative necrosis of the tumor with rupture of Bruch’s membrane and hemorrhage extending into the subretinal space (Fig. 5). At the edge of the tumor on the papillary side, a large choroidal artery and its branches were occluded by neoplastic cells (Fig. 6). The tumor did not extend into the subretinal space. Sections from more superior sectors revealed a hematoma centrally in the tumor and poor cytologic preservation of the adjacent tumor cells (Fig. 7). There was no inflammation within the tumor nor elsewhere in the specimen. Stains for iron showed slight to moderate staining around the hemorrhagic region of the mass and within some of the large macrophages in the overlying subretinal space.

Discussion

Despite recent advances in laboratory techniques such as the radioactive phosphorus uptake test and the radioactive chloroquine analog uptake test, the differential diagnosis of dark lesions at the macula remains difficult. Although ophthalmoscopic appearance and clinical course during a period of observation are the most valuable diagnostic aids, these aids are not completely reliable.

The present case is unusual in several respects. Although malignant melanomas of the choroid are found more frequently posterior to the equator, they are rarely present precisely at the macula. Subretinal and vitreous hemorrhages have been reported in association with malignant melanoma of the choroid, but such hemorrhages are found with large mushroom-shaped tumors which break through Bruch’s membrane and become engorged with blood as the neck of the tumor is constricted. These large tumors often have areas of necrosis. Hemorrhages are rare in small malignant melanomas of the choroid,
Fig. 1. Fundus drawing of the right eye showing the slightly elevated, subretinal macular mass and extensive retinal detachment due to subretinal hemorrhage.
Fig. 2. Overview of sectioned right eye showing choroidal melanoma and extensive hemorrhagic retinal detachment. Arrow indicates approximate location of the fovea (×3.5).

Fig. 3. Superotemporal calotte showing choroidal melanoma and a layer of degenerating blood on the retinal pigment epithelium. Central pale area of the tumor on the cut surface (arrow) corresponds to the region of coagulative necrosis (×5).

and their presence in association with small dark lesions at the macula is felt to be strong evidence against a diagnosis of malignant melanoma and to be indicative of hemorrhage disciform macular degeneration. A sudden change in the appearance of a small macular lesion, such as subretinal hemorrhage, is believed to weigh against a diagnosis of malignant melanoma. Exudative and serous detachments of the retina, with or without retinal holes or tears, have been reported with underlying melanomas of the choroid. However, extensive hemorrhagic detachment, as in this case, has not been previously reported.

The deterioration of central vision one year prior to enucleation probably coincides with the onset of malignant change and growth of the tumor. But, despite the relatively long period of observation and the impression that epithelioid tumors usually have a rapid growth rate and early extraocular extension or metastases, none of these factors appear to apply in this case. Although ocular inflammation is often a clinical and pathologic finding in choroidal melanomas with necrosis, this case is exceptional in that ocular inflammation was not noted clinically and inflammation did not accompany the coagulative necrosis demonstrated pathologically.

Infarction of the tumor was related to neoplastic invasion and occlusion of a large
Fig. 4. Histopathologic features of the choroidal melanoma in the viable area. The tumor consists exclusively of nodules of epithelioid cells with scattered pigmentation (hematoxylin and eosin, ×300).

Fig. 5. Region of ischemic infarction of the choroidal melanoma. Centrally, there is coagulative necrosis with rupture of Bruch's membrane and layering of extravasated blood in the subretinal space (hematoxylin and eosin, ×26).

choroidal artery which—judging from the size of the infarct—served a large part of the melanoma. Presumably, the occlusion occurred suddenly and thus precluded the development of collateral vascular supply. Hayreh11 recently emphasized the clinical importance of occlusive disorders of the posterior ciliary arteries and the physiologic function of the short posterior ciliary arteries as end-arteries in man. Although recent trends in the field of encology have emphasized immunopathologic causes for tumor necrosis,10 this case of vascular occlusion through neoplastic invasion should

Fig. 6. Margin of the melanoma (on the papillary side) showing invasion and occlusion of a large choroidal artery by neoplastic cells (arrow) (hematoxylin and eosin, ×500).

Fig. 7. Choroidal melanoma in the temporal paramacular region. Centrally, there is a large hematoma. The long posterior ciliary nerve is surrounded by viable tumor cells (hematoxylin and eosin, ×26).
remind us of this simple but sometimes neglected cause of tumor necrosis.

REFERENCES


