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Abstract

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PMID: 6345935 [PubMed - indexed for MEDLINE]
Malignant Melanoma in a Blind Eye

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A 62-year-old man developed a malignant melanoma in his left eye that had been blind due to trauma for 35 years. This case illustrates the difficulties in detection and early diagnosis of occult malignant melanoma of the uveal tract in phthisic globes. The necessity for long-term follow-up of posttraumatic eyes is stressed.

KEY WORDS: malignant melanoma, melanoma of eye, melanoma of blind eye

INTRODUCTION
Malignant melanomas arising from the uveal tract are the most frequent primary intraocular neoplasms in adults [1]. We are reporting a case of malignant melanoma arising in an eye that has been blind due to trauma for 35 years. Review of the literature and our case illustrates the clinical problems of such cases of melanoma in a blind eye which are quite different from the cases of melanoma arising in a functional eye.

CASE REPORT
A 62-year-old white man presented with a four-month history of progressive proptosis of the left eye associated with headache, eye pain, weight loss, and anorexia. Thirty-five years previously, the patient had been struck in the left eye by a nail while doing carpentry work, causing blindness. Because the injury was uncomplicated and not accompanied by pain, the eye was never removed.

Physical examination revealed a visual acuity of 20/20 in the right eye and no light perception in the left. The left eye was aphakic with a nonreactive pupil. The pupillary membrane could not be penetrated by indirect ophthalmoscopy. Intraocular pressure was 14.0 mmHg in both eyes. Eyelid function was normal bilaterally. Facial motor and sensory nerve function was intact. A firm mass was palpated about the left orbit.

A computed tomographic scan revealed a retroorbital mass within the muscle cone with questionable bony erosion nasally. The scan also revealed scleral opacity and calcification. An ultrasound study revealed extensive retinal detachment. Clinical diagnosis was exophthalmos with orbital tumor extending into retroorbital tissue.

The patient’s condition worsened rapidly with progressive enophthalmos and edema of the left orbital conjunctiva and ptosis of the left eyelid. He was begun on oral prednisone, resulting in a symptomatic improvement. Five days after admission, the patient underwent left orbital exenteration. At surgery, the tumor appeared to be encapsulated, however it was bound to the posterior aspect of the orbit. The tumor was freed from the orbital walls, the optic nerve was cut and the eye with the entire tumor was removed. No bony involvement was seen grossly. The patient did well postoperatively. Work-up for residual tumor and metastatic lesions was negative. He received a course of radiation therapy, 900 rads to the left orbit in three doses every other day over a period of six days.

PATHOLOGIC FINDINGS
Gross examination (Fig. 1) revealed a 2.0 cm in diameter globe and attached periorbital tissue. Posteriorly, there was a firm, well-circumscribed light-tan to brown tumor mass measuring 4.0 × 3.0 × 3.0 cm. The tumor was invading the retroorbital tissue, muscle, and fat. The retina and choroid within the globe appeared to be detached showing degeneration, old hemorrhage, fibrosis, and focal calcification. About 2 ml of thick brownish fluid was present on cut section of the globe. Serial sectioning of the tumor revealed a nodular firm consistency alternating with focal areas of softening, necrosis, and hemorrhage. The optic nerve was obliterated by the tumor mass.

Accepted for publication December 1, 1982.
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Histologically (Fig. 2), the lesion was a densely cellular neoplasm with cords and septa of fibrovascular tissue separating sheets of plump epithelioid cells intermingled with spindle shaped cells. Less solid areas showed a palisade arrangement of tumor cells oriented around blood vessels. Still other areas were composed of large bizarre multinucleated cells. The epithelioid component was characterized by cells with eosinophilic cytoplasm, round vesicular nuclei, and prominent central cosinophilic nucleoli. Many cells, both spindle and epithelioid, contained abundant granular brownish black pigment. The tumor was surrounded by a fibrous pseudocapsule intermingled with the scleral tissue of the orbit. Tumor cells were seen extensively invading the pseudocapsule.
There were also hemorrhagic necrotic foci with adjacent fibrosis. Tumor cells were invading the optic nerve, muscle, and fibroadipose tissue. Several sections demonstrated angioinvasion. This tumor pattern was diagnosed as an ocular malignant melanoma of mixed pattern arising from the choroid with extensive extracapsular invasion.

**DISCUSSION**

Ocular malignant melanomas arising in a blind eye present a difficult diagnostic problem for the ophthalmologist. The signs and symptoms present in a functional eye such as recent onset of blurred vision, intraocular hemorrhage, retinal detachment, or increased intraocular pressure may be absent or impossible to detect.

In two series of cases from the Armed Forces Institute of Pathology [2], one consisting of 969 eyes with opaque media and history of blindness for six months, 3.8% harbored unsuspected malignant melanoma. The other series consisted of 212 eyes with opaque media out of 1,000 cases of intraocular malignant melanoma of which 113 or 11.3% were unsuspected. Kirk and Patty reviewed 228 patients with choroidal melanoma and found that 24 or 10.5% were unsuspected cases [3].

As early as 1925, Neame and Khan determined that 10% of 402 eyes enucleated for glaucoma contained melanoma, 4% of which were blind eyes [4]. One of their cases was that of a patient who developed melanoma with extracocular extension 22 years after developing a posttraumatic cataract. A case report diagnosed as sarcoma in an atrophic blind eye 20 years after a herpetic infection may very well have been another case of malignant melanoma [5]. Microscopically, the tumor consisted of large sarcomatous cells with large pale nuclei and prominent nucleoli. Extraglacial extension as well as extensive tumor necrosis were noted. The cells were described as being nonpigmented with no further studies undertaken to characterize the tumor cells. On the basis of nonpigmentation, the possibility of malignant melanoma was simply disregarded despite highly suspicious clinical features and morphologic descriptions of the tumor favoring melanoma as the diagnosis.

Ocular malignant melanomas may arise from any site in the uveal tract, most frequently the choroid and ciliary body. Studies have been shown that most ocular melanomas probably develop in preexisting nevi [6-8]. Ocular melanomas have been originally categorized into six types by Callender [9]. With later modification, only three patterns are generally recognized, spindle A, spindle B, and epithelioid [10].

Prognosis depends upon cell type, size of the tumor, extraglacial extension, and in part site of origin in the uvea. Tumors arising in the iris, especially if they are of small size, are excisable and tend to behave in a benign fashion [10].

In regard to cell type, tumors comprised of spindle A cells are now considered essentially benign, reflected by their benign histologic features. In general, tumors composed of spindle cells (spindle A or B or pure spindle B) carry a better prognosis than those mixed with an epithelioid component. Pure epithelioid tumors have the worst prognosis [1].

Controversy regarding survival before and after enucleation questions any beneficial effect in resecting small intraocular melanomas. Survival statistics on patients with uveal melanomas indicates a low mortality rate prior to enucleation and a significant increase in mortality rate postenucleation [12]. This, at least, infers that operative manipulation may induce metastasis from a small lesion which might otherwise be treated on a more conservative basis.

Our case illustrates typical features of intraocular malignant melanoma arising in a blind eye. The tumor type was a mixed pattern of spindle and epithelioid cells accompanied by tumor necrosis and hemorrhage. Prognosis of our patient based on cell type was worsened by the advanced degree of extraglacial extension and evidence of angioinvasion. Exenteration of the orbit appeared as the only choice of therapy followed by postoperative radiation.

There are no clear-cut or consistent methods for adequate detection of choroidal melanomas, despite the relatively significant risk of such a tumor arising in a blind eye. Careful examination of the blind eye and retroorbital tissues especially for increased intraocular pressure is the best approach. Although in our case, intraglacial pressure was not elevated, occult melanomas are likely to produce symptomless glaucoma as the only significant finding in an opaque functionless eye. Eye pain as illustrated in our case may not occur until late in the course of the disease, indicating significant extraglacial extension.

**ACKNOWLEDGMENTS**

The authors thank Ms. Karen Dunn for excellent secretarial assistance.

**REFERENCES**
