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Location:	Basel, Switzerland
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Case number:	E 7919/24
Material Submitted:	Digital slide

Sudden pain and blindness in one eye

Clinical history:

A 41-year-old female presented with pain and progressive vision loss in the right eye over a three-month period, culminating in complete loss of light perception. Initially, the patient experienced only mild discomfort with dry eye symptoms. However, three days prior to presentation, she developed severe headaches, prompting her hospital visit.

Ophthalmic examination revealed right-sided exotropia with ptosis and exophthalmos. The conjunctiva demonstrated pronounced vascular injection. Anterior segment evaluation showed rubeosisiridis with a moderately dilated, non-reactive pupil. The lens was not visualizable, and fundus examination was not possible. Intraocular pressure was markedly elevated at 69 mmHg. During the examination, the patient experienced a seizure, necessitating postponement of further ophthalmic assessment and initiation of a comprehensive systemic workup.

Diagnostic imaging included magnetic resonance imaging, which revealed an intraocular tumor. Ocular ultrasonography then showed a characteristic "collar button" configuration. Based on these findings, enucleation was performed.

Ocular pathology:

Macroscopic examination:

The enucleated globe measured 25 × 25 × 23 mm with a corneal diameter of 18 × 12 mm. Sectioning revealed a dark red, polypoid lesion attached at the level of the optic nerve, measuring 15 × 13 × 12 mm. The lens was posteriorly displaced.

Microscopic examination:

The cornea remained intact. The iris demonstrated neovascularization. The retina was absent. The posterior segment contained regressively altered tumor tissue with faintly discernible spindle cells and melanin pigmentation. Immunohistochemical analysis showed strong positivity for HMB45 and partial positivity for Melan A and S100. The optic nerve contained melanin pigment deposits; however, immunohistochemical examination and deeper sectioning failed to demonstrate infiltrating tumor cells in this region.

Diagnosis:

Right eye enucleation specimen containing a regressively altered choroidal melanoma with associated secondary glaucoma and lens luxation.

Discussion:

Uveal melanoma represents the most common primary intraocular malignancy in adults, arising from the melanocytes of the iris, ciliary body, or choroid.[1] While typically diagnosed between 45-80 years of age,[2] our patient's presentation at 41 years is somewhat unusual. The clinical manifestations of uveal melanoma are variable, ranging from blurred vision and visual field defects to complete vision loss in advanced cases. Many patients remain asymptomatic until the tumor reaches sufficient size to compromise vision or cause retinal detachment.[3]

The management of uveal melanoma includes radiotherapy, enucleation, or local resection, depending on tumor size, location, and patient factors.[4] In this case, the advanced nature of the disease with secondary complications necessitated enucleation. Following surgery, staging revealed no evidence of metastatic disease. While adjuvant radiotherapy and surveillance were recommended, the patient declined further treatment or follow-up examinations.

Two notable complications in this case merit discussion. First, the lens luxation, although more commonly associated with melanomas of the ciliary body,[6] occurred despite the predominantly posterior (choroidal) location of the tumor. Second, the development of secondary glaucoma, characterized by dramatically elevated intraocular pressure (69 mmHg), likely resulted from neovascularization of the iris (rubeosisiridis) with consequent angle closure. Secondary glaucoma in the setting of uveal melanoma is associated with increased risk of metastasis and poorer prognosis.[7]

This case highlights the importance of considering uveal melanoma in the differential diagnosis of unilateral visual loss, particularly when accompanied by elevated intraocular pressure and anterior segment abnormalities. It also demonstrates the potential for serious local complications, including secondary glaucoma and lens displacement, which may significantly impact visual outcomes and management decisions.

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