EOPS Meeting 2025

Date of meeting	Basel, June 11 – June 14
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Case number:	24-1155
Material distributed	scanned glass slide

Ring Melanoma Presenting as Secondary Glaucoma

Clinical History

A 77-year-old male presented with acute pain in the left eye (OS) and progressive decline in visual acuity. On admission, visual acuity in the OS was reduced to counting fingers. Intraocular pressure (IOP) was markedly elevated in the OS (50 mmHg) compared to the right eye (OD) (11 mmHg). Slit-lamp examination of the OS revealed conjunctival injection, corneal edema, and discrete episcleral pigmentation. An initial diagnosis of anterior uveitis was considered. The patient was started on topical corticosteroids for inflammation control and received topical and systemic antiglaucomatous therapy to lower the IOP.

Despite treatment, IOP in the OS could only be reduced to a maximum of 25 mmHg following intravenous acetazolamide and local therapy. Ultrasonography also disclosed a 5×8 mm hyperechoic mass in contact with the ciliary body, lens, and iris, located in the temporal inferior quadrant. After regression of the corneal edema, fundoscopy revealed a very peripheral mass located behind the iris, between the 3 and 5 o'clock positions, raising suspicion of malignancy. Gonioscopy revealed a more pigmented chamber angle compared to the fellow eye.

Although the presentation initially mimicked anterior uveitis (i.e. masquerade syndrome), comprehensive investigation led to the diagnosis of a ring melanoma of the OS, with involvement of the anterior chamber angle and resulting in refractory unilateral secondary glaucoma. Due to the diffuse tumor infiltration in the chamber angle, enucleation was performed. The patient gave us informed consent to cultivate patient derived organoids (PDO) from his tumor.

Ocular Pathology

Gross examination.

Left eye measuring 24 × 26 × 26 mm. The cornea is clear, and the lens is slightly opacified. A pigmented ciliary body tumor is observed between 3:00 and 5:00 o'clock. The iris is darkly pigmented, with circular pigmentation in the chamber angle and pigment deposits on the posterior lens surface. Resection of three vortex veins and the optic nerve was performed.

Light microscopy

The cornea is structurally normal but shows a slightly reduced endothelial cell count. In the periphery, retrocorneal pigmented cells are present. The chamber angle is infiltrated by pigmented cells, with some areas containing solid nests of neoplastic cells. A tumor originating from the ciliary body is identified. The lesion measures 6 × 4 mm and infiltrates the ciliary body along with the iris root, breaching these structures and extending into the chamber angle. The tumor is composed primarily of Spindle B cells, which are elongated and display prominent nucleoli. Approximately two mitoses per high-power field are observed. Toward the iris root, the tumor cells exhibit increased pigmentation. In the chamber angle, infiltrating tumor cells and pigmented macrophages are also present. The neoplasm extends through the chamber angle, reaching Schlemm's canal and follows emissary channels. Pigmented cells are also found in the vitreous, behind the lens. The choroid, retina, and optic nerve are unremarkable. The sclera has normal thickness.

Immunohistochemistry.
Tumor cells are positive for Melan A, including the tumor segment that spreads into the chamber angle, anterior iris surface, trabecular meshwork, along Schlemm's canal, and the emissary channels. CD68 stains scattered macrophages, some of them pigmented. Ki-67 stains only a few tumor cells, consistent with a rather low mitotic rate.

Diagnosis **Ring melanoma, spindle B cell type, arising from the ciliary body, with circular infiltration of the chamber angle, extending into Schlemm's canal.**

Discussion

Ring melanoma is a rare subtype of uveal melanoma characterized by its circumferential growth pattern. It typically involves more than 180° (or 6 clock hours) of the ciliary body and may affect the iris and anterior chamber angle. Importantly, it is frequently associated with secondary glaucoma, as seen in our patient.

The tumor often has subtle onset, so patients may remain asymptomatic for a considerable period. When initial visual symptoms appear, they are usually nonspecific, contributing to a delayed diagnosis. Some cases may present with unilateral glaucoma refractory to conventional therapy. The elevated intraocular pressure (IOP) can result from various mechanisms, including tumoral invasion of the iridocorneal angle, iris neovascularization, and anterior displacement of the lens-iris diaphragm.

Thus, despite being a rare entity, this intraocular tumor should be considered when a patient has persistent monocular high intraocular pressure that does not respond to standard therapy. Early diagnosis is critical, as patient prognosis largely depends on the extent of tumor growth, with enucleation being common due to late-stage diagnosis. Prognosis worsens with anterior extension of the tumor and invasion of the Schlemm canal, which facilitates metastatic dissemination.

Diagnostic investigation includes a combination of resources, such as transillumination, ultrasonography and gonioscopy. Gonioscopy, in particular, may reveal angle involvement and/or pigment deposition at this site. However, it may be significantly impaired by corneal edema as initially in our case.

It is important to avoid open intraocular procedures (e.g., glaucoma surgeries) in suspected or established cases of ring melanoma because such interventions are associated with increased risk of neoplastic dissemination. Systemic spread of uveal melanoma typically occurs hematogenously. However, open eye procedures could allow the tumor cells to reach the lymphatic drainage system, through the conjunctival lymphatic vessels. Also, by invasion of Schlemm's canal, the tumor can get access to the venous systems via collector channels.

In conclusion, while ring melanoma is rare, it has significant implications due to its subtle presentation, diagnostic complexity, strong association with secondary glaucoma, and considerable intra- and extraocular extension. Our case emphasizes the importance of suspecting an intraocular neoplasm in cases of persistent unilateral elevation of IOP. The aggressive local and systemic spread of ring melanoma highlights the need for early detection and careful management.

References

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