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Intraocular Medulloepithelioma in a 15-Year-Old: A Rare Presentation

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Intraocular Medulloepithelioma in a 15-Year-Old: A Rare Presentation

Abstract

BACKGROUND: A 15-year old male presented with decrease in vision for the past 2 years. He was referred for a potential tumor due to an obstructed view of the right eye posterior segment. The patient was in good systemic health with no current medications.

FINDINGS: Best-corrected visual acuities were 20/63 OD and 20/16 OS through -4.00 DS OD and -4.25 DS OS. A dull fundus reflex was noted OD. Pupils were normal with no RAPD and biomicroscopy showed no cells or flare. IOP was 11 mm Hg OD and 12 mm Hg OS. The nasal quadrant of the right eye pupil failed to dilate completely. Fundus examination OD revealed diffuse vitreous haze and a large exudative retinal detachment due to a choroidal lesion spanning from 1 to 5 o'clock, sparing the macula.

ASSESSMENT: B-scan ultrasound showed a circumferential retinochoroidal mass with no calcification or excavation of underlying ocular structures. The dimensions of the tumor were 21.7mm × 21.2mm × 12.5mm. Ultrasound biomicroscopy showed anterior iris displacement from 3 to 5 o'clock. A large homogeneous mass containing multiple cystic spaces was noted. There was the presence of a lental apposition to the mass, retrolental cyclitic membrane. This atypical presentation led to the finding of a ciliary body tumor supplied by sentinel vessels extending to the retina causing exudative retinal detachment. The patient was referred for chest CT to rule out pleuropulmonary blastoma. MRI results showed hypointense signal in T2 weighted imaging with respect to the vitreous. Based on the findings we suspected this case to be an amelanotic melanoma or medulloepithelioma.

MANAGEMENT: The patient was referred to an ocular oncologist for fine needle aspiration biopsy under general anesthesia. Histopathology reports confirmed the presence of intraocular medulloepithelioma. Since the tumor base was too large for globe salvage and too thick for radioactive plaque, enucleation was performed and a vicryl mesh wrapped orbital ball implant was inserted.

Degree Type

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Degree Name

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James Kundart

Keywords

medulloepithelioma, oncology, exudative retinal detachment, ophthalmic imaging

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Intraocular Medulloepithelioma in a 15-Year-Old: A Rare Presentation

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Introduction

Intraocular medulloepithelioma is a congenital tumor of the ciliary epithelium that typically presents during the first decade of life. The histologic diagnosis is based on characteristic ribbons of pseudostratified neuroepithelium admixed with loose mesenchymal tissue rich in hyaluronic acid, vaguely resembling developing retina and vitreous. More than a third of medulloepitheliomas contain heteroplasic tissue, which in some cases makes up most of the tumor [1].

Case Report

A 15-year old Asian-Indian male presented with decrease in vision for the past 2 years. He was referred for a potential tumor due to an obstructed view of the right eye posterior segment. The patient was in good systemic health with no current medications.

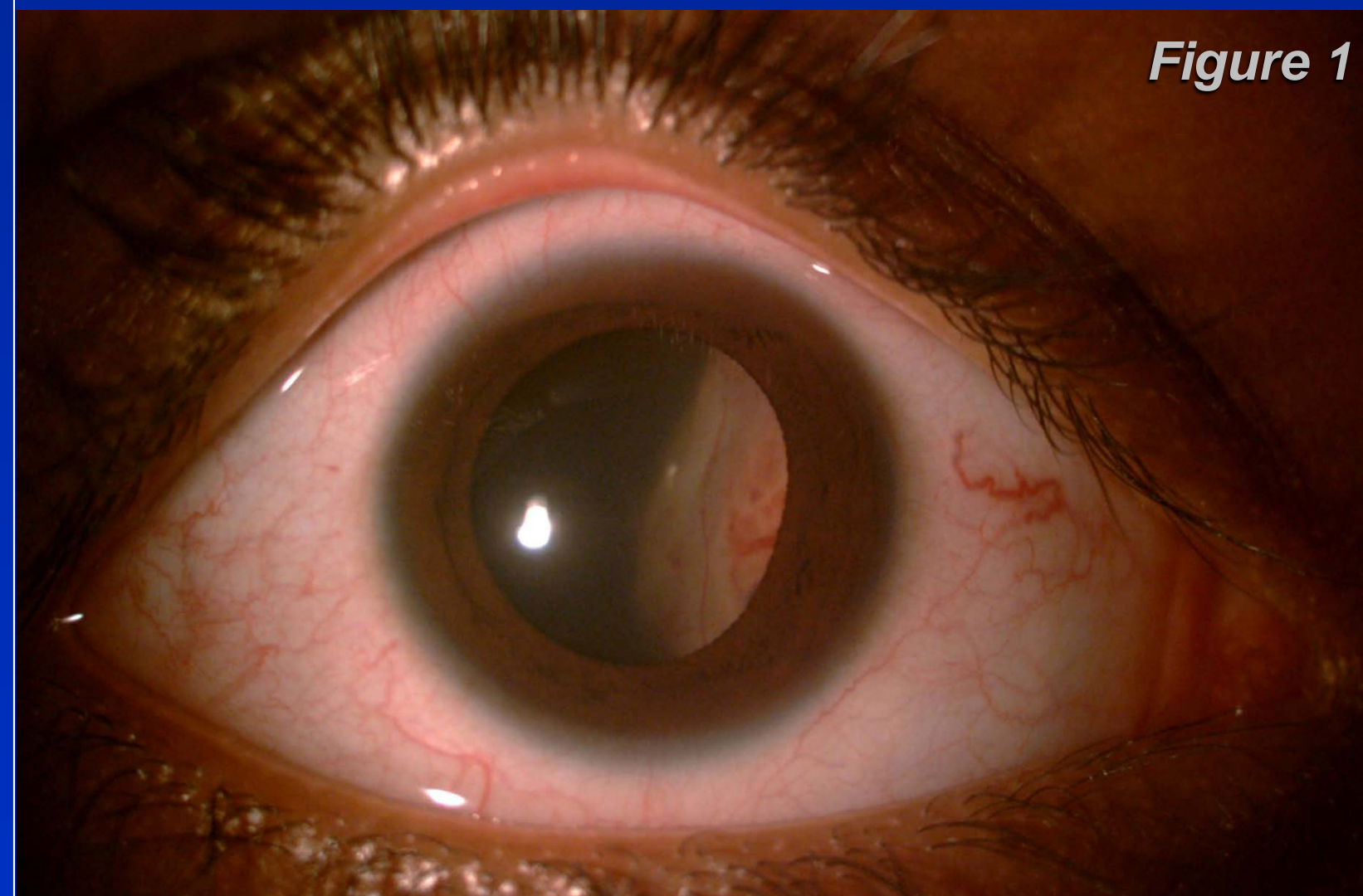


Figure 1

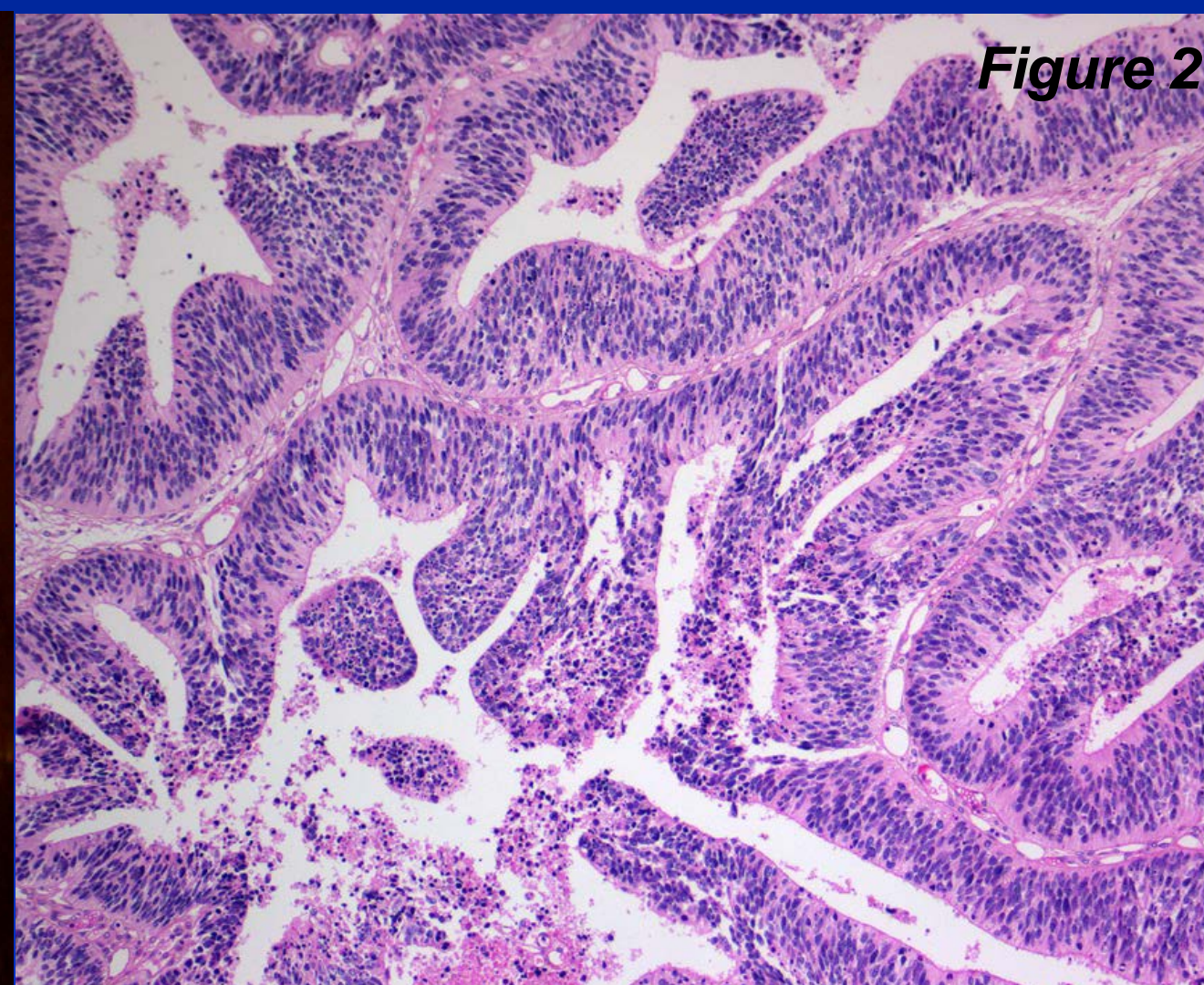


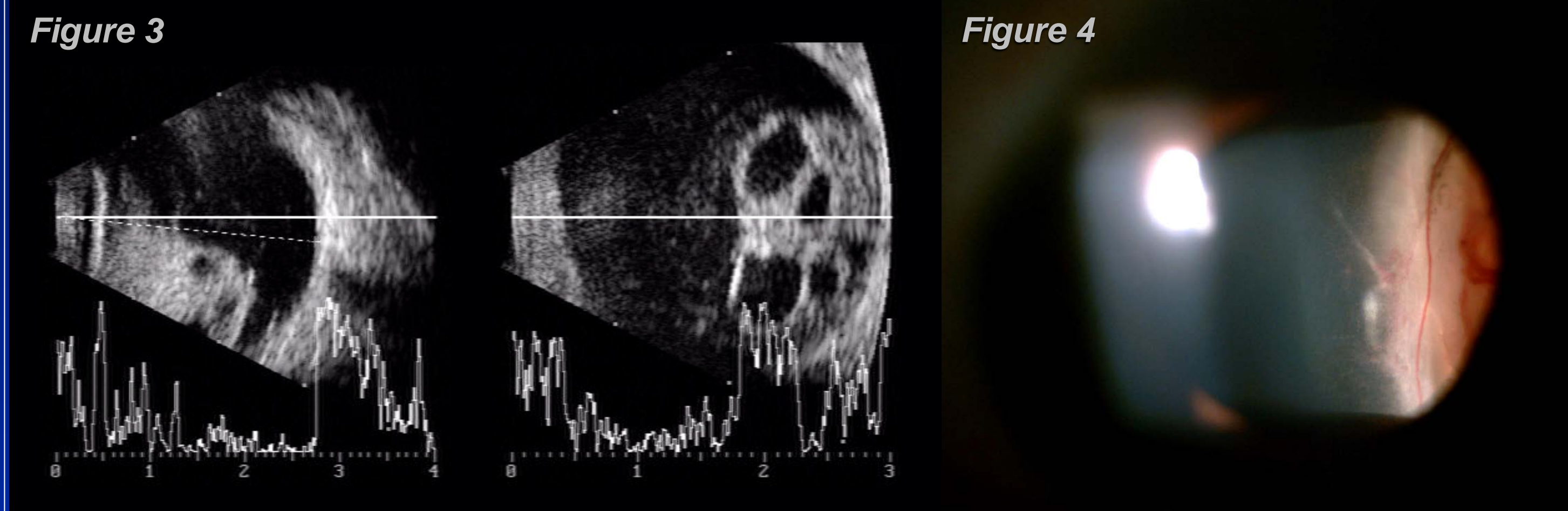
Figure 2

Findings

Best-corrected visual acuities were 20/63 OD and 20/16 OS through -4.00 DS OD and -4.25 DS OS. A dull fundus reflex was noted OD. Pupils were normal with no RAPD and biomicroscopy showed no cells or flare. IOP was 11 mm Hg OD and 12 mm Hg OS. The nasal quadrant of the right eye pupil failed to dilate completely (Figures 1, 4, 6). Fundus examination OD revealed diffuse vitreous haze and a large exudative retinal detachment due to a choroidal lesion spanning from 1 to 5 o'clock, sparing the macula.

Assessment

B-scan ultrasound showed a circumferential retinochoroidal mass with no calcification or excavation of underlying ocular structures (Figure 3). The dimensions of the tumor were 21.7mm X 21.2mm X 12.5mm.



Ultrasound biomicroscopy showed anterior iris displacement from 3 to 5 o'clock. A large homogeneous mass containing multiple cystic spaces was noted. There was the presence of a lental apposition to the mass, retrolental cyclitic membrane (Figure 5). OS was normal.



This atypical presentation led to the finding of a ciliary body tumor supplied by sentinel vessels extending to the retina causing exudative retinal detachment. The patient was referred for chest CT to rule out pleuropulmonary blastoma. MRI results showed hypointense signal in T2 weighted imaging with respect to the vitreous. Based on the findings we suspected this case to be an amelanotic melanoma or medulloepithelioma.

Management

The patient was referred to an ocular oncologist for fine needle aspiration biopsy under general anesthesia. Histopathology reports [2] confirmed the presence of intraocular medulloepithelioma (Figure 2). Since the tumor base was too large for globe salvage (~21mm) and too thick for radioactive plaque (12.5mm), enucleation was performed and a vicryl mesh wrapped orbital ball implant was inserted.

Discussion

This case demonstrates the importance of multimodal imaging in the diagnosis of intraocular medulloepithelioma. Clinicians should be aware of the distinct clinical onset and morphological characteristics of this tumor. Histopathology helps in confirming the pathogenic entity of this disease. Early diagnosis is crucial for preventing secondary ocular complications and globe salvage.

Clinical Pearls

- Manifests during the first decade of life
- Should be considered in the differential diagnosis of leukocoria
- White or gray translucent mass arising from the ciliary body
- Presence of cysts within the tumor, anterior chamber, or vitreous cavity
- Iris neovascularization, lens coloboma, sectoral or total cataract
- Other findings include a cyclitic neoplastic membrane, uveitis, hyphema, exudative retinal detachment, and vitreous hemorrhage

References

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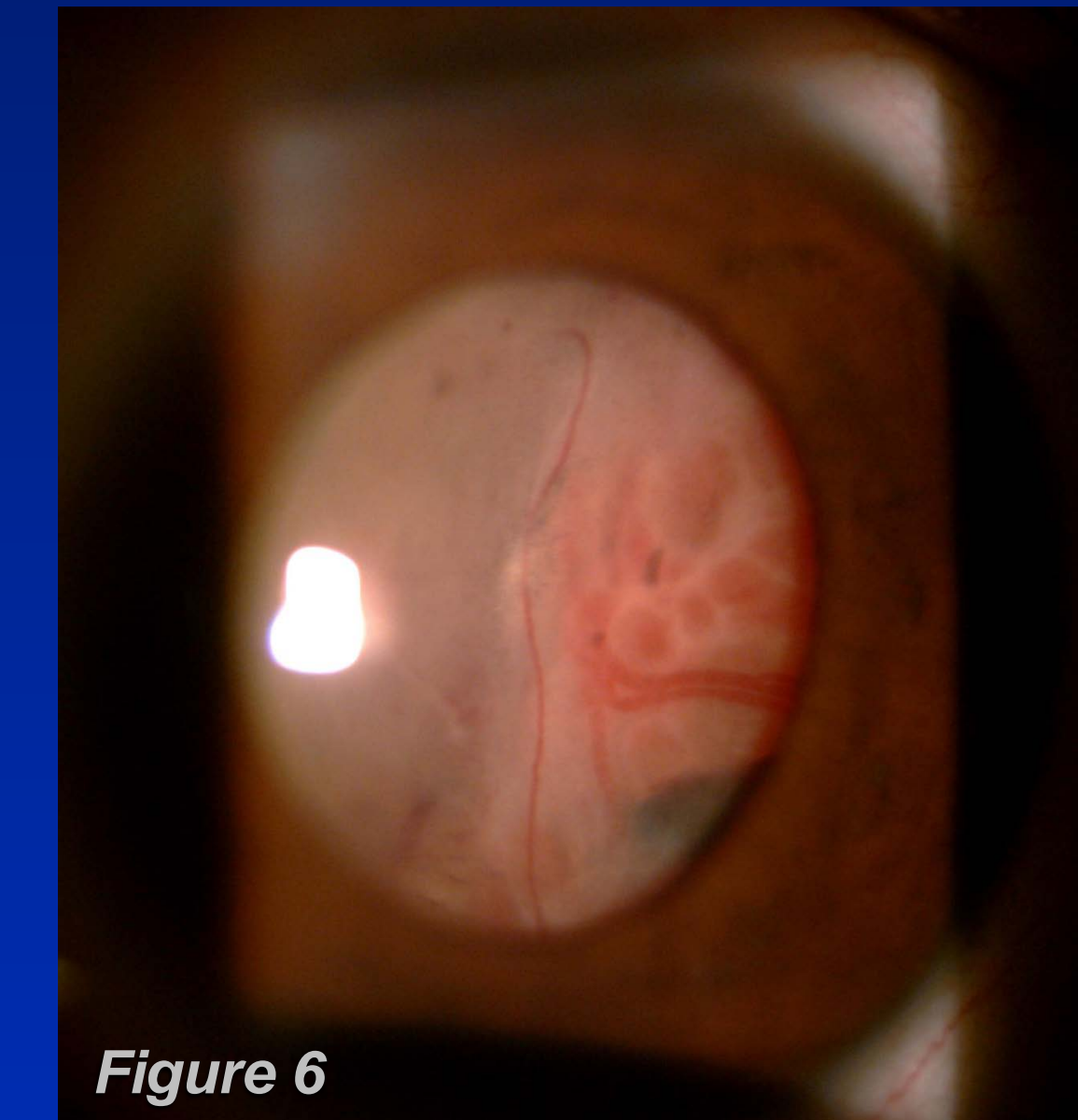


Figure 6