Fuchs Uveitis Syndrome Masquerading as "Phakic UGH"

Himani Akula BS[1], Redion Petrela MD[1], Robert Swan MD[1]

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Introduction

Fuchs Uveitis Syndrome (FUS), also known as Fuchs Heterochromic Iridocyclitis, is a rare form of chronic anterior uveitis that typically presents with unilateral heterochromia, low-grade iridocyclitis, and keratic precipitates.[1] This is a case of atypical FUS in the setting of congenital rubella which initially presented as uveitis, elevated IOP, and hyphema, suggestive of bilateral "phakic UGH syndrome"

Case Presentation

- 54F with history of congenital rubella presents with left eye pain, flashes, and severe blurriness
- Pt reports 30-year history of acute uveitis flare-ups and recently diagnosed glaucoma, taking brimonidine "intermittently"
- Initial exam showing PSC cataracts, elevated IOP, 2+ cell, TIDs, and smattered RBCs on gonioscopy (Figure 1).
- Initial presentation consistent with "phakic UGH syndrome". Differential included viral infection, Posner-Schlossman, chronic uveitis with damage to TM. Started on PF qid, brimonidine tid, xalatan qhs. AC tap to lower IOP and send for viral PCR
- IOP controlled. Subsequent dilated exam showing dense vitritis. Patient remained on long term steroid drops with improvement in inflammation.
- Cataracts progressed to 3+ NS and 3+ PSC. Underwent bilateral CEIOL with positive Amsler sign, suggestive of FUS. Rubella RNA was identified in the eye.
- Predforte reduced to PRN for symptomatic relief. Most recent visit VA was 20/20 OU with quiet AC and resolution of vitreous cellular reaction. Patient had not taken predforte for 4 months at time of final visit.

Exam Findings

Visual Acuity – Snellen

OD: 20/20	OS: 20/70
With glasses	OS: 20/40-2

Tonometry

OD: 37 OS: 48

Slit Lamp Exam

	Right	Left
Lids/Lashes	Normal	Normal
Conjunctiva/Sclera	White and quiet	White and quiet
Cornea	Clear, no KP 2+central guttata	Clear, no KP, +MCE
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Iris	subtle mid TIDS inf>sup	patchy TIDS inf>sup
Lens	Trace NS, central opacity	trace NS, central opacity
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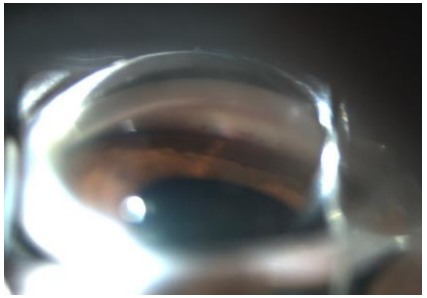

Figure 1: Gonioscopy showing RBCs at the inferior angle

This study was funded in part by unrestricted grants from Research to Prevent Blindness, Inc. New York, New York and Lions District 20-Y1, Syracuse, New York. No other significant financial interests or relationships to disclosure

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Discussion

- This case presents a diagnostic dilemma in which presenting symptoms were suggestive of bilateral UGH syndrome in a phakic patient.
- UGH Syndrome is typically seen in pseudophakia with AC or sulcus lens and is characterized by uveitis, glaucoma, and hyphema. Mechanical trauma by a mispositioned or subluxated IOL irritates the iris, leading to chronic inflammation, recurrent hyphema, and raised IOP.[2]
- A similar phenotype to UGH syndrome was seen in our patient; however it was observed bilaterally, and the patient was phakic.
- FUS is typically uniocular and is thought to be a result of viral-induced mild intraocular inflammatory response that may result in secondary glaucoma and cataracts.[3] It has been associated with HSV, CMV, ocular toxoplasmosis, and rubella and can be seen bilaterally in 10-20% of cases.[4]
- Chronic inflammation may cause trabecular meshwork obstruction, limiting aqueous humor drainage, increasing IOP, and resulting in glaucoma. Chronic inflammation may weaken blood vessels, increasing risk of rupture resulting in hyphema and further IOP increase.[5]
- Spontaneous hyphema may occur in severe disease due to iris rubeosis at the level of the trabecular meshwork.[5] This may present as Amsler sign during cataract surgery. Thus, bilateral Fuchs Uveitis may explain the "Phakic UGH" phenotype.
- In this patient, the diagnosis was unclear until cataract surgery, when Amsler sign was observed. Rubella RNA later identified in the eye further reinforced the diagnosis of FUS.
- Treatment of FUS includes a short course of corticosteroids, preferably topical, during flares and treatment for secondary complications like glaucoma and cataracts. Continuous monitoring is recommended. Long-term anti-inflammatory therapy is not indicated.[1]
- A complete history and eye exam is essential but may not be sufficient to distinguish between specific syndromes and/or diseases that have similar presentations. Thus, patients should be treated with personalized plans with regular monitoring and care. adjustment.

References

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- 54F with history of congenital rubella presents with left eye pain, flashes, and severe blurriness
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- This case presents a diagnostic dilemma in which presenting symptoms were suggestive of bilateral UGH syndrome in a phakic patient.
- UGH Syndrome is typically seen in pseudophakia with AC or sulcus lens and is characterized by uveitis, glaucoma, and hyphema. Mechanical trauma by a mispositioned or subluxated IOL irritates the iris, leading to chronic inflammation, recurrent hyphema, and raised IOP.[2]
- A similar phenotype to UGH syndrome was seen in our patient; however it was observed bilaterally, and the patient was phakic.
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Treatment

Treatment of FUS includes a short course of corticosteroids, preferably topical, during flares and treatment for secondary complications like glaucoma and cataracts. Continuous monitoring is recommended. Long-term anti-inflammatory therapy is not indicated.[1]

Conclusion

A complete history and eye exam is essential but may not be sufficient to distinguish between specific syndromes and/or diseases that have similar presentations. Thus, patients should be treated with personalized plans with regular monitoring and care. adjustment.

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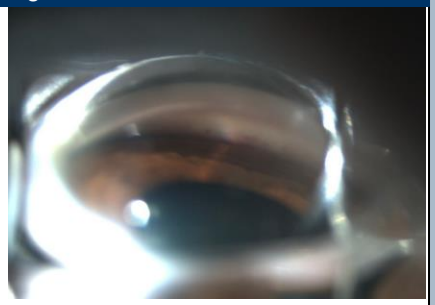

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Discussion

- This case presents a diagnostic dilemma in which presenting symptoms were suggestive of bilateral UGH syndrome in a phakic patient.
- UGH Syndrome is a sequela of pseudophakic IOLs and is characterized by uveitis, glaucoma, and hyphema. It is not a diagnosis that can be made in a phakic patient, however a similar phenotype was seen in our patient.
- Fuchs Uveitis is typically uniocular and is the result of a viral particle induced mild intraocular inflammatory response that may result in secondary glaucoma and cataracts (3). Pathophysiology involves…
- Spontaneous hyphema may occur in severe disease due to iris rubeosis at the level of the trabecular meshwork (3). This may present as Amsler sign during cataract surgery. Thus, bilateral Fuchs Uveitis may explain the "Phakic UGH" phenotype.
- Congenital Rubella is frequently associated with Fuchs Heterochromic Iridocyclitis or Fuchs' Uveitis.
- Rare occurrence of bilateral Fuchs uveitis have been seen with toxoplasmosis (6), Usher's Syndrome retinitis pigmentosa (5), and unknown etiologies (4). Infectious agents like rubella, herpes simplex, toxoplasmosis, and parasites have been known to induce Fuchs' Uveitis
- In bilateral disease, the classic heterochromia may be subtle or absent
- In this patient, Amsler sign during cataract surgery and Rubella RNA in eye solidified the diagnosis of Fuchs Uveitis Syndrome.

- involves…

Conclusion

The initial presentation of bilateral uveitis, high intraocular pressure, and hyphema alongside other associated signs like a transillumination defects and glaucoma initially presented as "Phakic UGH" phenotype. However, bilateral manifestation of Fuchs Iridocyclitis may also explain symptoms.

The previously listed symptoms, Amsler Sign during cataract surgery, and Rubella RNA particles in the eye cultures is most consistent with bilateral Fuchs Iridocyclitis most likely due to patient's congenital exposure to Rubella.

A complete history and eye exam is essential, but may not be sufficient to distinguish between specific syndromes and/or diseases that have similar presentations. Thus, patients should be treated with personalized plans with regular monitoring and care adjustment.

References

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