

Fuchs Heterochromic Iridocyclitis and Posner-Schlossman Syndrome

Early detection and diagnosis are essential to properly managing these diseases.

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Fuchs heterochromic iridocyclitis (FHI) and Posner-Schlossman syndrome (PSS) are closely related inflammatory causes of glaucoma. This article describes specific etiopathologies and general strategies for managing these conditions.

FUCHS HETEROCHROMIC IRIDOCYCLITIS Presentation

Initially described in 1906 by Austrian ophthalmologist Ernst Fuchs, FHI generally presents as a mild unilateral inflammation of the anterior chamber with stellate keratic precipitates and iris heterochromia with prominent iris and angle vessels secondary to iris atrophy that represents about 2% to 7% of cases of anterior uveitis.¹ In rare cases, FHI may present bilaterally with occasional evidence of “snowbanking” at the pars plana. Patients usually have symptomatic vision loss secondary to cataract formation and tend not to have significant inflammation on examination. Elevated IOP is not a consistent phenomenon but can occur secondary to peripheral anterior synechiae, trabecular scarring, rubeosis, lens-induced angle closure, recurrent spontaneous hyphema, and steroid-induced ocular hypertension.

Etiology

Historical etiological theories for FHI included sympathetic denervation, which could account for iris

hypochromia, and infectious etiologies such as herpes simplex virus, toxoplasma, and cytomegalovirus (CMV). Evidence increasingly suggests, however, an association between the rubella virus and FHI. In a study looking at aqueous humor antibody index and the presence of viral genome by polymerase chain reaction (PCR), rubella antibodies were detected in almost all eyes with FHI, and the rubella genome was found in almost half of the patients who were younger than 40 years old.² Cellular component analysis in FHI reveals a predominance of T-lymphocyte markers CD3 and CD4, suggestive of viral etiopathogenesis.³ CMV was also identified by PCR testing in a large number of eyes with clinical evidence of FHI.⁴ Thus, a viral etiology would explain FHI’s general unresponsiveness to topical steroid therapy, which in many cases is unnecessary and can have untoward side effects.

Evidence has also suggested that the incidence of FHI has decreased since the introduction of the measles, mumps, and rubella vaccinations.⁵ Ocular manifestations of congenital rubella syndrome include congenital cataracts, glaucoma, microphthalmia, and pigmentary retinal changes, and this occurs in infants born to mothers infected in the first trimester. Rubella antibodies are seen in children infected after the first trimester, indirectly suggesting a late-trimester infection that may manifest as FHI.⁶

FHI and Glaucoma

Glaucoma occurs in 10% of patients with FHI at presentation, with an incidence risk of 0.5% per year.⁷ It is important that ophthalmologists watch for a rise in IOP and the formation of peripheral anterior synechiae with chronic damage to the trabecular meshwork, especially after cataract surgery, which can lead to intraoperative bleeding from angle vessels.

Management

Medical management is a reasonable but generally futile option, with failure reported in more than 70% of patients. Although trabeculectomy with antimetabolites is successful in 60% to 70% of patients 2 years postoperatively,⁸ glaucoma drainage implants have over a 90% success rate 1 year postoperatively and a 50% success rate after 4 years postoperatively.⁹ Our preference is to implant a pediatric Ahmed Glaucoma Valve (New World Medical, Inc.) with an intracameral injection of dexamethasone (Decadron; Merck & Co., Inc.). Cataract surgery generally results in good visual outcomes, but it may be associated with hyphema, uveitis, a small pupil, and zonular dehiscence.

POSNER-SCHLOSSMAN SYNDROME

Presentation

Initially described by Posner and Schlossman in 1948, PSS or glaucomatocyclitic crisis present with recurrent unilateral attacks of significantly elevated IOP and minimal conjunctival injection, epithelial corneal edema, and anterior chamber reaction in the form of small keratic precipitates. Iris atrophy and heterochromia are frequently seen, whereas posterior synechiae typically are not. Patients have open angles with normal IOP in between attacks, which usually last a few hours to a few weeks.

Etiology

The various etiological theories proposed include autonomic, vascular endothelial, and autoimmune dysfunction, but the infectious theory implicating CMV seems to have the most evidence.¹⁰ Other organisms implicated in PSS include *Helicobacter pylori*, varicella-zoster virus, and herpes simplex virus.

Chee et al found in a cohort study that one-quarter of patients with anterior uveitis tested positive for CMV by aqueous humor PCR testing and that a large majority of these patients had clinical evidence of PSS.¹¹ The likelihood of being positive by PCR testing was greater during an active episode of PSS. Oral antiviral therapy reduced the frequency of attacks and other sequelae in these patients. Although the seroprevalence of CMV is high worldwide,¹² the low frequency of PSS

Weigh in on this topic now!



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1. How frequently do you encounter Fuchs heterochromic iridocyclitis or Posner-Schlossman syndrome?

- Often
- Sometimes
- Rarely
- Never

2. With which treatment modality have you had the most success in cases of Fuchs heterochromic iridocyclitis?

- Medical therapy
- Trabeculectomy with antimetabolites
- Glaucoma drainage implants
- Cataract surgery

3. With which treatment modality have you had the most success in cases of Posner-Schlossman syndrome?

- Topical IOP-lowering medications
- Steroids
- Cycloplegic agents
- Filtering surgery

(1.9/100,000)¹³ may suggest that other genetic (eg, HLA-Bw54)¹⁴ or coinfectious factors (eg, *Helicobacter pylori*) may be necessary for clinical manifestation.

PSS and Glaucoma

In a study by Jap et al, about 25% of patients with PSS had evidence of glaucoma, and those with 10 or more years of clinical activity had a 2.8 times higher risk of developing glaucoma.¹⁵ In a small series of patients, changes to the optic nerve head were shown to be transient in the majority of cases.¹⁶

Management

The treatment of PSS includes topical IOP-lowering medications, steroids, and cycloplegic agents. Filtering surgery is successful in preventing IOP spikes and for long-term IOP control.¹⁷ We start with medical therapy, which includes prednisolone 1% and topical drops to lower IOP and reserve incisional surgery for refractory cases.

LINKS BETWEEN THE TWO DISEASES

Both FHI and PSS have common etiopathogenic and clinical factors. A study that compared patients with FHI and PSS found that over 50% of PSS patients and about

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40% of FHI patients were positive for CMV.⁴ Patients with presumed FHI who tested positive for CMV were generally older men with nodular endothelial lesions. There were no clinical differences between CMV positive and negative patients with presumed PSS.

CONCLUSION

Both FHI and PSS are challenging problems. Careful clinical examination followed by appropriate ancillary testing and treatment is an effective strategy by which to manage FHI and PSS. Clinicians should carefully consider and administer antiviral therapy in eyes with recalcitrant uveitis. Early detection and diagnosis with appropriate observation are a must in combating these diseases. ■

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- Wakefield D, Chang JH. Epidemiology of uveitis. *Int Ophthalmol Clin*. 2005;45(2):1-13.
- Quentin CD, Reiber H. Fuchs heterochromic cyclitis: rubella virus antibodies and genome in aqueous humor. *Am J Ophthalmol*. 2004;138(1):46-54.
- Gordon L. Fuch's heterochromic cyclitis: new clues regarding pathogenesis. *Am J Ophthalmol*. 2004;138(1):133-134.
- Chee SP, Jap A. Presumed Fuchs heterochromic iridocyclitis and Posner-Schlossman syndrome: comparison of cytomegalovirus-positive and negative eyes. *Am J Ophthalmol*. 2008;146(6):883-889.
- Birnbaum AD, Tessler HH, Schultz KL, et al. Epidemiologic relationship between Fuchs heterochromic iridocyclitis and the United States rubella vaccination program. *Am J Ophthalmol*. 2007;144(3):424-428.
- Jones NP. Glaucoma in Fuchs' heterochromic uveitis: aetiology, management and outcome. *Eye (Lond)*. 1991;5(6):662-667.
- Rothova A. The riddle of fuchs heterochromic uveitis. *Am J Ophthalmol*. 2007;144(3):447-448.
- La Hey E, de Vries J, Langerhorst CT, et al. Treatment and prognosis of secondary glaucoma in Fuchs' heterochromic iridocyclitis. *Am J Ophthalmol*. 1993;116(3):327-340.
- Da Mata A, Burk SE, Netland PA, et al. Management of uveitic glaucoma with Ahmed Glaucoma Valve implantation. *Ophthalmology*. 1999;106(11):2168-2172.
- Takusagawa HL, Liu Y, Wiggins JL. Infectious theories of Posner-Schlossman syndrome. *Int Ophthalmol Clin*. 2011;51(4):105-115.
- Chee SP, Bacsal K, Jap A, et al. Clinical features of cytomegalovirus anterior uveitis in immunocompetent patients. *Am J Ophthalmol*. 2008;145(5):834-840.
- Cannon MJ, Schmid DS, Hyde TB. Review of cytomegalovirus seroprevalence and demographic characteristics associated with infection. *Rev Med Virol*. 2010;20(4):202-213.
- Paivonsalo-Hietanen T, Tuominen J, Vaahtoranta-Lehtonen H, Saari KM. Incidence and prevalence of different uveitis entities in Finland. *Acta Ophthalmol Scand*. 1997;75(1):76-81.
- Hirose S, Ohno S, Matsuda H. HLA-Bw54 and glaucomatocyclitic crisis. *Arch Ophthalmol*. 1985;103(12):1837-1839.
- Jap A, Sivakumar M, Chee SP. Is Posner-Schlossman syndrome benign? *Ophthalmology*. 2001;108(5):913-918.
- Dinakaran S, Kayarkar V. Trabeculectomy in the management of Posner-Schlossman syndrome. *Ophthalmic Surg Lasers*. 2002;33(4):321-322.
- Darchuk V, Sampaioles J, Mato L, et al. Optic nerve head behavior in Posner-Schlossman syndrome. *Int Ophthalmol*. 2001;23(4-6):373-379.