Pars Planitis

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Case Report

A ten-year-old Hispanic boy from Mexico was referred for one month history of blurry vision and floaters in the right eye.

His past medical history was significant for recurrent group A Streptococcus infections of pharynx with elevated titer of antistreptolysin O antibodies, recurrent mouth sores, and headaches.

We found visual acuities of 20/50 and 20/25, and intraocular pressures of 19 and 15 mmHg right and left eye, respectively. The slit lamp examination revealed 2+ cells in the anterior chamber OD and 1.5+ cells in the anterior chamber OS. Ophthalmoscopy disclosed 3+ cells in the vitreous OD, 2+ cells in the vitreous OS, and an exudate with blurry margins on pars plana between 3 and 9 o,clock. Hence, the diagnosis of PP in both eyes was made.

The patient received transseptal injection of Kenalog (triamcinolone 40 mg/ml) in the right eye and a serologic work-up and ENT consult were requested at the initial visit. The patient returned for a follow-up visit two weeks later. The visual acuity in the right eye improved to 20/40 and the eye examination revealed diminished, but still active PP in the right eye. There was no change in visual acuity or activity of PP in the left eye. The report from the ENT specialist confirmed that the patient is a carrier of group A Streptococcus and the patient was placed on oral Augmentin. The serologic studies showed mildly elevated level of angiotensin converting enzyme.

Because the patient and his family were not able to spend an extended period of time in the United States or to regularly return for regular follow-up visits that would be required if a long-term systemic immunomodulatory therapy for active bilateral PP is initiated, pars plana vitrectomy (PPV) was suggested to the patient,s parents. After discussing risks and benefits of PPV the parents decided to move along with surgery. Hence, the patient underwent PPV with endolaser and cryopexy first in the right and three month later in the left eye. The patient returned to Mexico on no systemic or topical medication one moth after each surgery.

There was no evidence of active PP in either eye on follow-up visits as late as two years after PPV. The patient,s visual acuities were 20/25 OD and 20/20 OS.

Definition

Pars planitis (PP) is defined as peripheral vitritis with exudate overlying peripheral retina, ora serrata, or pars plana ciliaris.

Pars planitis is typically a sign of intermediate uveitis, a more broad clinical entity defined as inflammation of anterior vitreous with or without presence of pars plana exudate. Some authors use PP and intermediate uveitis as synonym. The following review focuses on PP proposed by the above definition rather than on intermediate uveitis per se.

History

The clinical entity similar to PP was first described by Fuchs in 1908. A more detailed description of the disease and its association with peripheral retinal vasculopathy and exudation at pars plana were reported by Schepens in 1950. The term PP was first used by Welch in 1960.

Epidemiology

The average age at the diagnosis of PP is 26 years. The diagnosis is, however, reported in individuals as young as 5 years and as old as 50 years of age. The disease is more common in women with male to female ratio of 2:3.

Pathogenesis

The etiology and pathogenesis of PP remain unknown. Main histocompatibility gene typing showed that 64% of individuals with PP are carriers of HLA-DR 15 gene, 57% of HLA-DR 51 gene, and 29% are carriers of HLA-DR 17 gene suggesting that genetic predisposition to PP exists.

Clinical Manifestation

In addition to exudate on pars plana, peripheral retina, or ora serrata, the hallmark of PP, anterior uveitis is observed in 49% and vasculitis in the periphery of retina in 17 % and disc edema in 17 % of patients. The course of PP can be complicated by cystoid macular edema (68%), cataract formation (57%), epiretinal membrane (40%), retinal detachment (9%), retinal neovascularization (8%). Less frequent complications of PP include vitreous

hemorrhage (3-5%), band keratopathy (2%), and posterior synechiae (2%). Furthermore, some individuals may develop cyclitic membrane (**Figure 1**) and/or autoimmune endotheliopathy.

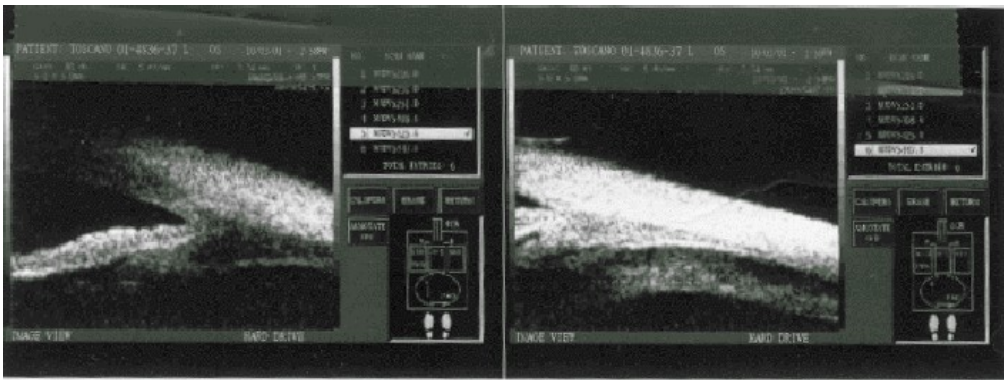

Figure 1 Cyclitic membrane in a patient with pars planitis who presented with hypotony.

An association between PP and a systemic disease is reported in 45% of patients. The remaining 55% of cases are not associated with presence of a systemic disorder. Multiple sclerosis and sarcoidosis represent systemic diseases most commonly associated with PP. The onset of multiple sclerosis in patients with PP is reported between 17 years prior to diagnosis of PP and 7 years after the diagnosis of PP. Interestingly, 11% of patients with PP have a first degree relative with multiple sclerosis. Sarcoidosis in patients with PP is typically diagnosed in the interval between the onset of PP and 5 years after the onset of PP.

Scanty reports describe PP in patients with retinitis pigmentosa, cat scratch disease or Ebstein-Barr virus infection.

Therapy

We currently follow a step-ladder approach in treatment of patients with PP. Periocular steroid injections are the first choice. Patients who continue having PP or those who develop a recurrence of PP are indicated to systemic therapy. Non-steroidal anti-inflammatory drugs are typically employed initially. If the activity of PP is not controlled with non-steroidal anti-inflammatory drugs immunosuppressive therapy is indicated.

Our experience and experience of others suggest that cryopexy alone or in combination with pars plana vitrectomy may result in induction of durable remission of PP (this was also the case of our patient described above).

We are currently conducting a prospective randomized trial comparing safety and efficacy of early pars plana vitrectomy to early immunosuppressive therapy in patients with PP.

Prognosis

The extraordinary high frequency of complications of PP (see above) that ultimately lead to irreversible damage to the retina and/or optic nerve and impairment of vision and blindness strongly speak for early aggressive therapeutic intervention. It is noteworthy that some authors recommend to initiate treatment of PP if the visual acuity is worse than 20/40. We do not share this opinion unlike many others and initiate therapy as soon as the diagnosis is established. We believe that this approach is associated with better prognosis and less vision rubbing complications.