Pars Planitis (Idiopathic Intermediate Uveitis of the Pars Planitis Type)

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Introduction

Intermediate uveitis (IU) is the general terminology for inflammation localized predominantly to the vitreous, ciliary body, and peripheral retina that may or may not be associated with an infectious agent or systemic disease. Pars planitis (PP) is a subset of IU where the cause is idiopathic, and clinical findings include snowbank or snowball formation. Such nomenclature is an important distinction as PP is a diagnosis of exclusion that is made only after a thorough investigation into potential infectious or autoimmune causes has been conducted.

As the majority of pediatric patients with uveitis have an indeterminate cause, PP is most often diagnosed in this patient population. Due to the wide spectrum of clinical findings, the reported incidence and prevalence of this disease is highly variable. The Northern California Epidemiology of Uveitis Study found the incidence to be 1.5 per 100,000 population years, while a study from a tertiary referral center, which tends to see more severe cases of uveitis, reported that 15.4% of uveitis patients had PP. The gender distribution of PP remains uncertain. The prevalence of childhood-onset PP appears to be lowest in patients of Hispanic background; however, further studies are necessary to elucidate these findings.

Since PP is a diagnosis of exclusion, the underlying mechanism for this disease is not well understood. Studies have suggested a genetic predisposition, and various haplotypes such as HLA-DR2, HLA-DR15, HLA-B51, and HLA-DRB1*0802 have been associated with the disease. There is a strong association with the development of multiple sclerosis in patients with PP, and individuals with either the HLA-DR2 or HLA-DR15 haplotype showed the highest association.

Case 1: Pars Planitis in a Young Lady Requiring Antitubercular Therapy

A 32-year-old Asian Indian female presented with diminution of vision in both eyes for the past 2 months. Her best-corrected visual acuity (BCVA) was 20/100 and 20/125 in right (OD) and left eyes (OS), respectively. Slit-lamp examination revealed flare in the anterior chamber (no cells) and plenty of vitreous cells in both eyes (OU) on retro-illumination. Fundus examination revealed a hazy media (grade 2) along with significant vitritis. The foveal reflex was blunted. There were a few snow-balls in the inferior periphery of OD (Fig. 1). Fluorescein angiography (FA) revealed presence of significant early macular leakage which increased in the late phase (petalloid pattern) suggestive of cystoid macular edema (CME). In addition, there was significant disc leakage in the late phase which was suggestive of optic nerve inflammation. Peripheral vascular leakage

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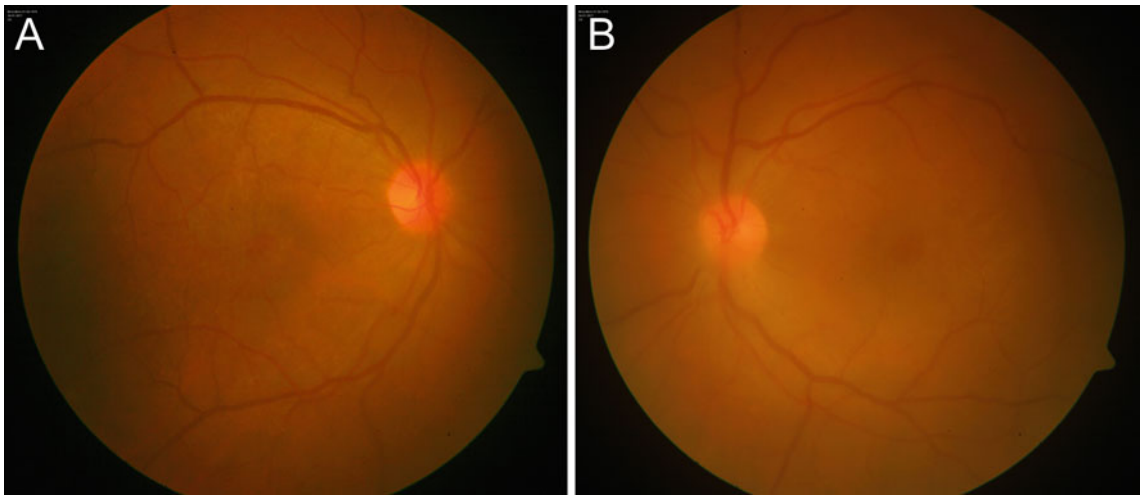

Fig. 1 Fundus photography of patient #1 (Case 1) bilateral pars planitis. The fundus photographs of the right (**a**) and left eye (**b**) shows presence of vitritis, mild disc swelling (especially in the right eye), and dull foveal reflex

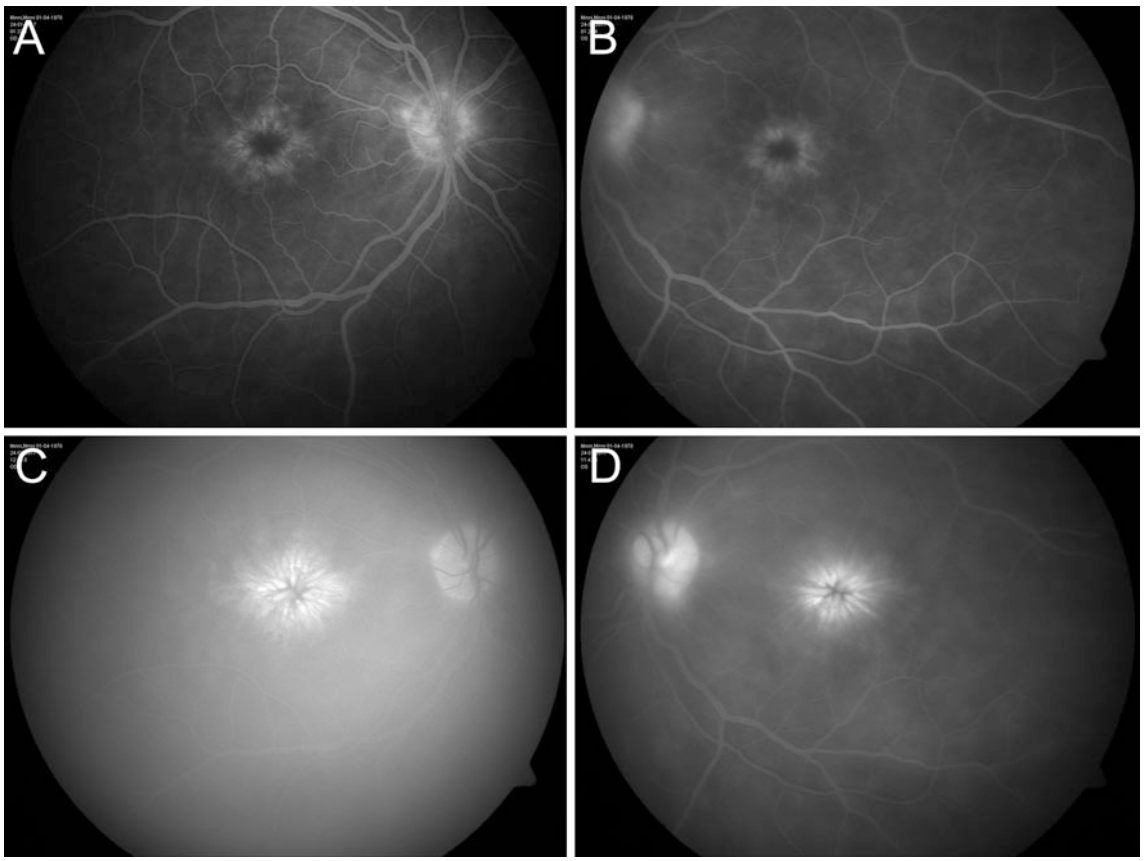

Fig. 2 The FA in the early phase (**a**) and (**b**) shows presence of early disc hyperfluorescence and perifoveal hyperfluorescence especially in the right eye indicative of cystoid macular edema. The late frames (**c**) and (**d**) show presence of significant disc hyperfluorescence in both eyes and macular leakage suggestive of cystoid macular edema in both eyes

was also seen in the peripheral sweeps of the FA (Fig. 2). The patient was diagnosed with intermediate uveitis. Her Mantoux test was highly positive (>30 mm induration at 48 h) and she had a positive QuantiFERON Gold TB® test.

In view of the significant inflammation, macular edema, and optic disc leakage, she was initiated on 1 mg/kg oral corticosteroids (60 mg/day oral prednisone) along with anti-tubercular therapy (consisting of isoniazid, ethambutol,

rifampicin, and pyrazinamide). Due to persistent inflammation and intolerance to oral corticosteroids, oral azathioprine was added 1 month after antitubercular therapy as a steroid-sparing agent. 3 months after therapy, her BCVA was recorded as 20/30 OU. Her inflammation has significantly subsided and she is on revised therapy with antitubercular agents along with 2 mg/kg/day oral azathioprine.

Case 2: Macular Edema in a Patient with Pars Planitis

A 43-year-old Asian Indian male patient presented with diminution of vision in the right eye for the past 3 months. His BCVA was 20/80 in OD. Clinical examination using slit-lamp biomicroscopy revealed presence of 1+ vitreous cells along with a hazy media. Fundus examination showed a dull foveal reflex (Fig. 3). FA revealed the presence of disc hyperfluorescence along with perifoveal leakage suggestive of CME (Fig. 3). Optical coherence tomography (OCT) of OD revealed presence of intraretinal cystoid spaces confirming the presence of CME.

The patient was thoroughly investigated for the etiology of the uveitis. However, all the tests were negative. A diagnosis of PP was considered. The patient had only unilateral involvement with macular edema and was counseled to receive an injection of intravitreal dexamethasone implant (Ozurdex®). One month after the injection, the patient had marked resolution in the macular leakage on FA (Fig. 4) and resolution of the intraretinal cystoid spaces on OCT. His BCVA improved from 20/80 to 20/25 in OD.

Key Points

- While PP has a variable natural course and prognosis, most cases show progressive worsening of inflammation and tissue damage in the absence of therapy. It is therefore imperative to strategically initiate a system of escalating anti-inflammatory therapy with/without immunosuppressive agents based upon the clinical findings and disease response.
- Depending upon the severity of vitreous inflammation, some patients may develop blinding sequelae and complications such as tractional RD and optic disc edema.
- In general, the disease prognosis is worse in patients diagnosed at a younger age. Given the severe and debilitating nature of PP, patients should be thoroughly evaluated for other causes of IU such as multiple sclerosis, and prompt management with corticosteroids, immunosuppressive

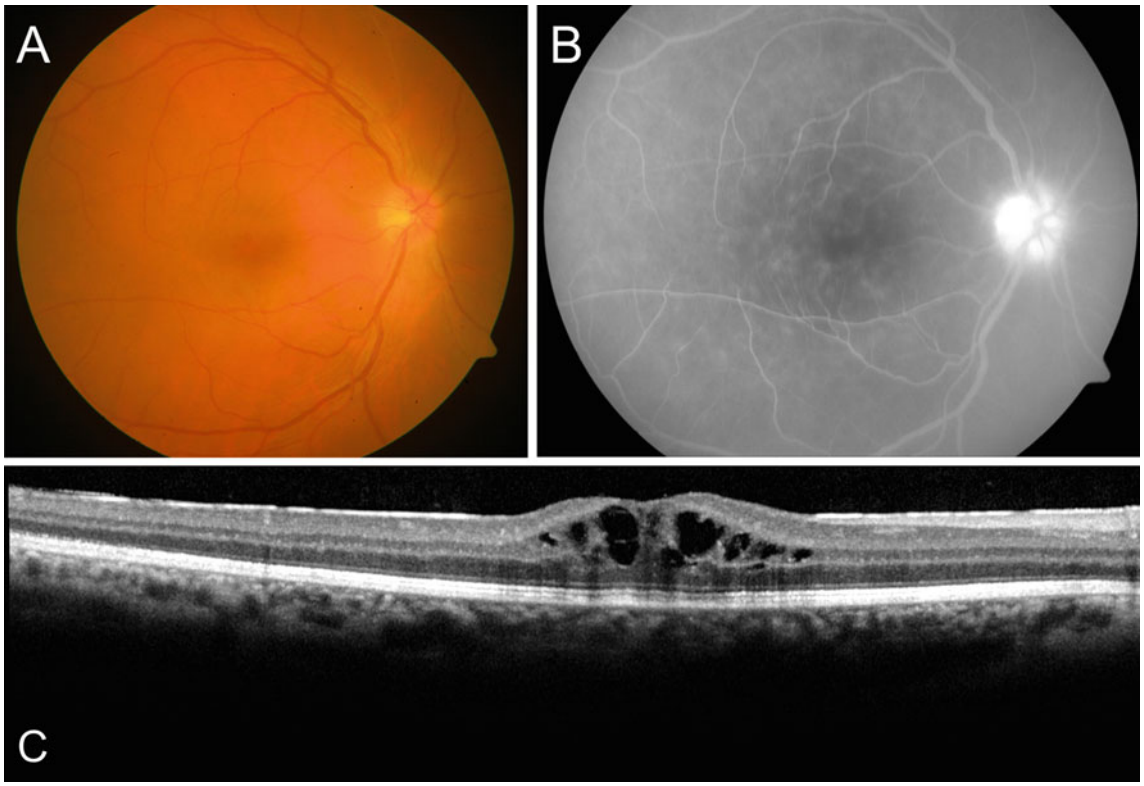

Fig. 3 Fundus photograph (**a**) of patient #2 (Case 2) with unilateral pars planitis involving the right eye (OD) shows media haze due to vitritis and a dull foveal reflex due to underlying macular edema. Fluorescein angiography (**b**) shows presence of perifoveal leakage suggestive of macular edema and disc hyperfluorescence. Optical coherence tomography of OD (**c**) shows presence of multiple cystoid spaces in OD along indicating cystoid macular edema

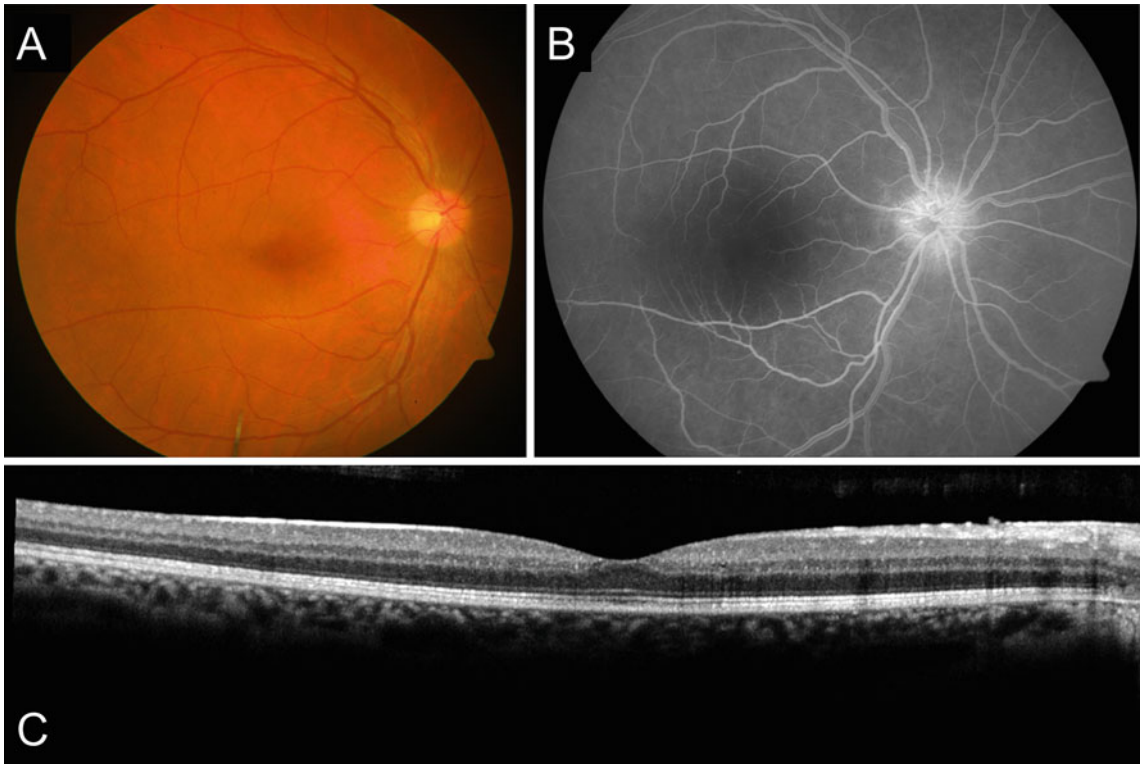

Fig. 4 Fundus photograph (**a**) and fluorescein angiography (**b**) of the same patient (patient #2, Fig. 3) after treatment with intravitreal injection of dexamethasone implant (Ozurdex ®) recorded after 1 month shows resolution of vitritis and perifoveal leakage on fluorescein angiography. The disc is also normal without hyperfluorescence seen in Fig. 3. Optical coherence tomography of OD (**c**) shows resolution of cystoid macular edema in OD seen in Fig. 3

agents, or biological agents should be initiated when indicated.

- Multimodal imaging is employed in the management of these patients to understand and determine the exact extent of tissue damage in eyes with pars planitis.

Suggested Reading

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