Intermediate Uveitis

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Case Presentation

A 32-year old woman presented with visual loss in her right eye on8/9/95. She was diagnosed with bilateral pars planitis with vitreoushemorrhage and inferior snowbank in the left eye in 1986. Subsequently she developed rhegmatogenous retinal detachment in that eye. Pars plana vitrecromy with endolaser panretinal photocoagulation (PRP) and scleral buckling OS was done. One year later she had cataract extraction with posterior chamber IOL implantation OS. Her right eye was treated with monthly periocular corticosteroid injections and peripheral cryotherapy. She was intolerant to Prednisone and discontinued Motrin after 2 months because "it didn't work". Review of systems revealed paresthesias in legs for the past 10 years, multiple allergies, arthritis, sinusitis, peptic ulcer and seizures since she had encephalitis in 1989.
VA was 20/70 OD and counting fingers from 2 feet OS. Intraocular pressures were 19 mmHg OD, 9 mmHg OS. Slit lamp examination revealed normal anterior segment findings in the right eye; a few small,round white keratic precipitates (KPs) inferiorly, 2+ cells and 1+ flare in the anterior chamber OS. There was a large superior peripheral iridectomy OS. PC IOL was coated with inflammatory cells and there was a dense posterior capsule opacification. Fundus examination of the right eye revealed 1+ vitreous cells, cystoid macular edema, mild optic disc edema and peripapillary edema which were confirmed by fluorescein angiography (Figure1).

Fig 1
There was a collagen band over the pars plana and pigmentary changes in the peripheral retina OD. There was no view due to hazy media in the left eye.
Laboratory work-up was non-conributory, including angiotensin coverting enzyme(ACE), serum lysozyme,PPD, chest X-ray, FTA-abs and MRI of the head.
Trans-septal(TS) Kenalog was administered in the right eye. Dolobid 500 mg twice a day and cyclosporin A (CSA) 200 mg/d was prescribed. She couldn't get CSA because the insurance company refused to pay for that; methotrexate (MTX) 7.5 mg/w was started in place of CSA. Topical Ocufen 4x1 OU, Pred-Forte 2x1 OD 4x1 OS was prescribed.
On follow-up she developed multiple recurrences in both eyes which were treated by TS Kenalog injections and increasing MTX dose. In flare-ups VA dropped to 20/40-20/50 and improved to 20/30 with treatment.On 4/2/96 Nd-YAG capsulotomy was done in the left eye. On 10/7/96 PPV and PC IOL explantation OS was done.There was inflammatory cells, debris and white, plaque-like posterior capsule opacification inferiorly. Because of this appearance possibility of chronic postoperative endophthalmitis was also considered. (**Figure 2**)

Fig 2
Culture of the vitreous specimen and IOL was negative for malignant cells and microorganisms. On her last examination on 4/8/97, she was on CSA 200mg/d (started one month previously) and MTX 17.5 mg/w.She was tolerating both drugs well and there was a marked improvement of symptoms and signs after addition of CSA to the drug regimen.VA was 20/25 OD, 20/400 OS. Anterior segment was clear in the right eye. There was 1+cells and 2+flare in the AC of the left eye. IOP were 20mmHg OD and 9mmHg OS.On fundus examination there was a mild CME and peripheral pigmentary changes in the retina of the right eye (Figures 3).

Fig 3

In the left eye there were 360 degrees scleral buckle, laser scars and gliotic membrane in the inferior peripheral retina. She was kept on the same medical therapy and PPV/Epiretinal

membrane (ERM) peeling was recommended for the left eye.
This case is an example of intermediate uveitis with many characteristic findings: long duration (>10 years), bilateral involvement with marked asymmetry, development of complications (e.g. cataract formation, retinal detachment), poor prognosis for IOL implantation; required removal of the IOL (perioperative inflammatory status and medical management seems to be inappropriate according to the history and signs in this case) and effectiveness of immunosuppressive therapy (CSA+MTX) and probably PPV, as well.

INTERMEDIATE UVEITIS

Intermediate uveitis is a common type of uveitis in children and young adults. It is one of four major categories of uveitis in the classification scheme proposed by the International Uveitis Study Group (IUSG)(1). This classification scheme subdivides intraocular inflammation into anterior, intermediate, posterior, and panuveitis, based on the principal anatomic site of infammation. The diagnosis of intermediate uveitis is made when intraocular inflammation primarily involves the vitreous, peripheral retina and pars plana ciliaris. It is notorious for its long duration; in fact, intermediate uveitis is the type of uveitis whose clinical duration is the longest (2). Although the majority of cases are idiophatic and the patients have no systemic disease, a significant association between intermediate uveitis and sarcoidosis, multiple sclerosis (MS) and Lyme disease is widely accepted (3,4).

History and Terminology

Many different names have been used to describe this uveitis entity. It was first described by Fuchs in 1908 as "chronic cyclitis" (5).The term "peripheral uveitis was used by Schepens in 1950 (6), "pars planitis" by Welch et al. in 1960 (7), "vitritis by Gass in 1968 (8), and finally "intermediate uveitis" is the term suggested by IUSG in 1987 (1). Use of the terms intermediate uveitis and pars planitis remains confusing. Most authors prefer "pars planitis" when there are pars plana exudates and collagen band (snowbank) over the pars plana. Snowbank is not required for the diagnosis of intermediate uveitis but it is associated with a worse prognosis (9).

Epidemiology

Intermediate uveitis accounts for approximately 10% of uveitis cases in a referral practice. Although the incidence of uveitis in children is low, intermediate uveitis may account for up to 20% of uveitis cases in children. Onset is typically between 2nd and 4th decades. Onset after 40 years of age is rare. The age of onset correlates inversely with the severity of expression: when symptomatic within the first decade of life the severity of inflammation, the resulting vitreous opacification, and the resistance to therapy are significantly greater than when the onset occurs in the second to fourth decades.Early onset is not common and is seen in about 10% of affected patients (10).
Bilaterality approaches 80% with long term follow-up; the severity may differ between the two eyes and it may be only unilateral. A study from Korea is atypical in that intermediate uveitis was unilateral in 101 of 107 cases (94.4%) (11).There appears to be no sex or race predilection. There are occasional cases of familial pars planitis (12).HLA-DR2 association with intermediate uveitis has been reported (13,14). HLA-DR2 has also been associated with multiple sclerosis which is of potential interest in that some patients with multiple sclerosis also have intermediate uveitis.

Clinical Features

Patients may be asymptomatic or more commonly complain of blurry vision and/or floaters. Onset is insidious which is in contrast to the acute onset seen in infectious causes of vitritis such as acute retinal necrosis or toxoplasmosis. There is usually no pain, photophobia or redness.

Signs

The eye is typically "white". There may be a mild anterior segment inflammation with a few small, round, white keratic precipitates (exceptions: in chidren, in MS, Lyme disease and sarcoidosis there is usually more severe inflammation in the anterior segment).Patients with intermediate uveitis associated with MS typically develop a granulomatous anterior uveitis with mutton-fat keratic precipitates which can mimic sarcoidosis (15,16,17).
Three to 9 percent of eyes with intermediate uveitis develop band keratopathy due to chronic anterior segment inflammation (18).
An autoimmune endotheliopathy in the inferior cornea similar to the phenomenon of corneal allograft rejection (i.e., a collection of distinct keratic precipitates arranged linearly at the junction of edematous and nonedematous inferior cornea) was described by Khoudadoust et al. in 4 of 10 patients with intermediate uveitis (19). This observation has not been noted as frequently by others (20).
Peripheral anterior synechia and posterior synechia occur uncommonly, usually in the setting of an acute onset or chronic anterior segment inflammation.
Vitritis is the most consistent sign of intermediate uveitis. It may become so dense as to obscure the retina entirely (21,22). These white cells are in both the anterior and posterior vitreous (diffuse clouding), have the appearance of "dust" at the slit lamp biomicrocopy and can be seen through the undilated pupil. Characteristic mobile, globular, yellow-white "snowballs" ("ants' eggs") can be seen in the inferior peripheral vitreous.They lie close to the retina, but are not in contact with it. They are not specific for pars planitis and may occur with any kind of inflammation of the peripheral fundus or with an extensive and diffuse uveitis (23). Later the vitreous shows degenerative changes with fiber-like cylinderic condensations of coarse vitreous strands. Posterior vitreous detachment (PVD) is common; PVD is uncommon in normal eyes of individuals under 40 yr of age. The hallmark of pars planitis are the white or yellowish-white pars plana exudates ("posterior hypopyon") and collagen band (snowbank) over the pars plana. These exudates are preretinal, peripheral, typically inferior but may also be superior or divided into multiple foci or extend 360 degrees over the entire pars plana (**Figure 5**).

Fig 5
This peripheral exudation and snowbanking are best seen either with the Goldmann three-mirror lens or with a 20+ diopter lens and scleral indentation.
Cystoid macular edema (CME) is the major cause of visual loss both at early onset and in the long term follow-up.It is confirmed by fluorescein angiography and it necessitates therapeutic intervention either at the onset or during the follow-up, usually with periocular corticosteroid injections. A case report of a patient with chronic intermediate uveitis and associated classic snowbanking (pars planitis) with severe CME, probably due to Lyme borreliosis is noteworthy. Despite a disease duration of 10 years the patient's ocular symptoms and CME, hence the visual acuity, responded promtly to intravenous ceftriaxone treatment (4). The amount of macular edema may not correlate with the degree of vitreous inflammation.
Optic nerve is usually normal but can be slightly hyperemic and swollen; severe optic disc swelling is unusual and should suggest syphilis or sarcoidosis. Peripheral retinal vasculitis, primarily involving venules is common. It is seen as sheathing and/or irregularity in caliber and can be subtle ophthalmoscopically; it is best appreciated by fluorescein angiography (**Figure 6**).

Course and

Prognosis

Most series describe a chronic course that may have periodic exacerbations and remissions (21). In the long term follow-up study by Smith et al.,it appeared that pars planitis ran three patterns of disease: (1) a self-limiting course characterized by gradual improvement without a single episode of exacerbation of the low grade activity (10%); (2) a prolonged course without exacerbations (59%); (3) a chronic smoldering course with one or more episodes of exacerbation (31%) (24).In one study (54 patients with idiopathic pars planitis, 108 eyes), the authors found a greater risk of retinal detachment in patients with significant cataract formation; periphlebitis at the time of diagnosis appeared to increase the risk of development of optic neuritis or MS. They also reported an overall favorable visual prognosis in patients with pars planitis(25).

Durations of active disease up to 15 to 20 years are not uncommon. Compared to other types of uveitis, the duration of intermediate uveitis is among the longest (2,21).
Study of advanced or chronic cases allows several conclusions (22). First, visual acuity is generally only mildly impaired at the time of initial presntation. Second, the prognosis for good vision is related to severity more than to duration, although both play a role. Third, macular disease (CME and post cystoid degeneration) is the most important visual prognostic factor. Fourth, the presence of a pars plana exudate is associated with worse prognosis.Lastly, although patients are occasionally counseled that their disease will "burn out" over time, permanent spontaneous resolution of intermediate uveitis rarely occurs; and vision loss occuring by the time the disease "burns out" is common.

Complications

In a series of 100 patients with a follow-up ranging from 4 years to more than 20 years (median and mean of 10.5 years), the complications of 182 eyes were studied. The most common complication was cataract formation (42%), then CME (28%), followed in order of decreasing incidence by band keratopathy, glaucoma, retinal detachment, retinoschisis, vitreous hemorrhage, "retinitis pigmentosa-like" changes, and dragged disc vessels (18). Neovascularization in the optic disc (NVD), elsewhere in the retina (NVE) or in the snowbank can occur and peripapillary subretinal neovascularization has also been reported (26,27).

Pathology

The pathologic and clinical literature to date suggests that the retinal venous system is the primary inflammatory focus (22,28,29). In all eyes examined there is prominant perivascular lymphocytic cuffing (perivasculitis) and mural infiltration of the retinal vessels (vasculitis), primarily involving the venous system (periphlebitis/phlebitis). Venous involvement is usually anterior to the equator.The uveal tract is typically free of inflammation.
Light microscopic examination of the vitreous "snowbank" reveals a collapsed, condensed vitreous base, blood vessels, scattered lymphocytes, spindle-shaped cells, and hyperplasia of nonpigmented ciliary body epithelium. Electron microscopy shows the proliferating cells in the collapsed viteous base to be fibrous astrocytes producing new large diameter collagen fibrils. The snowbank is in fact a fibroglial inflammatory aggregate, occasionally with a neovascular component. Electron microscopic examination also revealed high endothelial venules (HEVs) (29).These specialized venules are lined by plump endothelial cells that, on cross section protrude into the vessel lumen. HEV formation is a consequence of local T cell activation and T cell-derived cytokine production, particularly interferon gamma (IFN-g) triggered by antigen.

These are activated endothelial cells which express a variety of adhesion molecules and HLA classII molecules, not found on the flat, resting endothelial cells of ordinary venules. These molecules in turn play important roles in lymphocyte traffic and antigen presentation to T cells, respectively. It is suggested that cryotherapy is effective by eliminating these HEVs. Immunohistologic examination of vitreous cells identified T lymphocytes as the predominant vitreous cell in 15 eyes of 13 patients with intermediate uveitis (30). Macrophages were the second most common cell type; B lymphocytes were scarce.
Etiology of intermediate uveitis is unknown. The response of this disease to immunosuppressive therapy, familial clustering, and occasional association with other presumed autoimmune diseases such as multiple sclerosis combine to imply that autoimmunity plays a role in the pathogenesis of intermediate uveitis (22). The putative antigenic stimulus remains obscure. Intermediate uveitis may occur with spirochetal infections caused by Treponema pallidum or Borrelia burgdorferi (31,4). Any hypothesis regarding the etiology of intermediate uveitis should clarify the major clinicopathologic findings in retinal venous system and vitreous base.

Differential Diagnosis
· Toxocarisis
· Toxoplasmosis
· Acute retinal necrosis
· Lyme disease, syphilis
· Human T cell lymphotropic virus type1 (HTLV-1) ass. uveitis
· Whipple's disease (Tropheryma whippelii)
· Endogenous endophthalmitis (bacterial, fungal)
· Irvine-Gass syndrome
· Spillover from iridocyclitis (e.g. Fuchs' heterochromic uveitis)
· Multiple sclerosis
· Sarcoidosis
· Intraocular lymphoma
· Amyloidosis
· Inflammatory bowel disease
· Behcet's disease
· Intraocular foreign body

Differential diagnosis of intermediate uveitis is principally differential diagnosis of vitritis in children and young adults. As in any case of uveitis it is important to rule out infectious causes, as specific treatment and cure can be achieved and most patients with idiopathic intermediate uveitis need immunosuppressive therapy.
In children, toxocariasis is a major diagnostic consideration when dealing with unilateral intermediate uveitis. Patients with ocular toxocariasis are usually free of systemic findings. The characteristic unilateral chronic endophthalmitis with peripheral granuloma and tractional bands extending to the posterior pole (which does not necessarily develop in the inferior peripheral fundus) which can be confirmed by echography is distinct from intermediate uveitis.History of infected puppies or pica and serologic testing (ELISA) for Toxocara antigen can also be helpful. Toxoplasma retinochoroiditis is typically unilateral. Active toxoplasma lesions are round or oval, yellow-white, adjacent to a pigmented old scar or satellite lesions and are usually in the posterior retina. Peripheral lesions, probably due to toxoplasmosis have been described, including a wide ring-like lesion near the extreme periphery resembling the snowbanking seen in pars planitis (15,32). Active peripheral toxoplasmic retinochoroiditis may cause such a heavy vitreal reaction that the retinal lesion itself cannot be directly visualized ("headlight in the fog").There may be "spillover" anterior segment inflammation with small to medium-sized, round white or large, mutton-fat KPs in the cornea.It is usually acute in onset in contrast to the insidious onset in intermediate uveitis. Diffuse or segmental retinal vasculitis involving both arterioles and venules in the vicinity of, as well as remote to the lesion can be seen (Figure 7).

Fig 7

In contrast,in intermediate uveitis retinal venules anterior to equator are involved."Scaffolding" of vitreuos bands with round, yellow KP-like inflammatory cells, reminiscent of beads on a string may be seen.
Acute retinal necrosis (ARN) due to herpes viruses usually presents acutely in one eye (the second eye becomes involved in almost 1/3 of cases, usually within 1 to 6 weeks) with pain, redness and photophobia. Severe vitritis with dense white infiltrate in retinal periphery (necrotizing retinitis) can be confused with pars planitis. Acute onset,rapid progression of peripheral retinal necrosis (usually circumferentially, sometimes with finger-like projections posteriorly), and findings in the posterior retina such as optic neuropathy and vasculitis (mainly arteritis) can be helpful in differentiation from intermediate uveitis.In suspicious cases diagnostic vitrectomy should be considered.
The possibility of an occult intraocular foreign body must always be kept in mind when dealing with unilateral intermediate uveitis. A fibroglial membrane can form over a foreign body resting on the pars plana, obscuring it from view and simulating pars planitis.

Diagnostic Tests

There is no specific laboratory test that confirms the diagnosis of intermediate uveitis. Diagnostic testing is performed either to rule out common or treatable causes of vitreal inflammation, to determine the cause of decreased vision, or to guide treatment.
The initial laboratory ivestigation may include complete blood count (CBC) with differential, erythrocyte sedimentation rate (ESR), chest X-ray, serum angiotensin converting enzyme (ACE), PPD, FTA-abs, and Lyme disease antibody titers.Additional testing is performed based on the clinical circumstances, as in the case of suspected toxocariasis, toxoplasmosis, multiple sclerosis, or intraocular lymphoma.
Ultrasonography can be useful when visualization of the retina is obscured to determine the extend and density of vitreous involvement and retinal anatomic status, i.e., tractional or rhegmatogenous retinal detachment. Echographic studies showing a solid, highly reflective peripheral mass and a retinal fold (or tractional detachment) between the mass and the optic nerve are useful in confirming the diagnosis of toxocariasis.
Fluorescein angiography is the most useful test, both diagnostically and as a guide to treatment, and confirms CME,optic nerve involvement, peripheral retinal vasculitis, and rare complication of neovascularization.

Treatment

If a specific cause (such as Lyme disease or syphilis) can be identified, then treatment is directed against these disorders.
Most patients with idiopathic intermediate uveitis require therapeutic intervention at some point during the course of their disease. Topical corticosteroids are reserved for clinically important anterior segment inflammation, as they do little to decrease posterior segment inflammation and may cause or enhance well-known complications of cataract formation and glaucoma. In general, a stepwise approach to therapy in patients with intermediate uveitis is most appropriate:
1. Periocular steroid therapy (e.g. Kenalog, 40mg/ml) is appropriate first-line therapy for most patients. A series of three injections at two to 3-week intervals should disclose by the end of two months whether or not the patient will respond. Systemic corticosteroids are considered in patients who do not respond to periocular treatment or those who cannot tolerate the injections and/or in bilateral cases. Oral steroids are usually given initially in a dose of 1mg/kg/day for 2-3 weeks, and then slowly tapered by 10mg/day in succeeding weeks if the patient responds.
2. Patients who are unresponsive or intolerant to corticosteroid therapy may be treated by peripheral cryotherapy. Cryotherapy is applied to the area of snowbank and one probe width adjacent retina under direct observation by indirect ophthalmoscopy, using a single or double freeze-thaw technique.Periocular corticosteroids are administered at the conclusion of the

procedure. Retreatment may be required, which is generally delayed 3 to 4 months to allow time for the initial treatment response.
3. If the previous measures fail, immunosuppressive drugs such as cyclosporin, azathioprine, methotrexate, and cyclophosphamide may be used with careful monitoring of the efficacy and potential side effects. Combination therapy may be more revelant in this respect. Nonsteroidal antiinflammatory drugs (e.g. Diflunisal 500mg twice a day) and acetazolamide may be adjunctive drugs for maintenance and chronic CME therapy, respectively.
4. Pars plana vitrectomy is more commonly being used as a diagnostic and therapeutic modality in intermediate uveitis. It is indicated in the management of complications such as retinal detachment, vitreous hemorrhage, cataract formation (pars plana lensectomy/vitrectomy) and in cases refractory to medical therapy. Intraocular lens implantation in intermediate uveitis is controversial. If it is considered, ocular inflammation should be preferably "quiescent" for at least 3 months prior to surgery and kept in abeyance following surgery (33). Therapeutic PPV may be effective by clearing the vitreous from debris and possible "antigenic load" and by removing possible traction in the macula, thereby improving or stabilizing CME. In appropriate cases it may be considered before immunosuppressive therapy in the management of intermediate uveitis.
The goal of treatment in intermediate uveitis must be elimination of the inflammatory process; however, this rarely is achieved. More realistically, the ophthalmologist must strive to reduce the severity of the inflammatory process. A more satisfying therapeutic end point awaits definite delineation of etiologic agent(s) or process of this enigmatic disease (10).

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Intermediate Uveitis Review Questions

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1. Which of the followings about epidemiology of intermediate uveitis is true?

a) It is more common in elderly patients.

b) It is more common in children and young adults.

c) It is more common in Mediterranean countries.

d) It is more common in females.

2. Which is not one of the terms used in the literature for intermediate uveitis?

a) Peripheral uveitis.

b) Chronic cyclitis.

c) Pars planitis.

d) Recurrent multifocal choroiditis.

3. Which one is true about intermediate uveitis?

a) Onset is usually acute.

b) Visual acuity is usually less than 20/40 at onset.

c) It is unilateral in 80% of cases.

d) Cystoid macular edema is common and is the major cause of visual loss.

4. Which is one of the characteristic findings in intermediate uveitis?

a) Severe anterior segment inflammation with posterior synechia.

b) Severe optic disc swelling.

c) Severe dust-like vitritis.

d) Significant arteritis in the retina.

5. Which one of the following signs has not been reported in idiopathic intermediate uveitis?

a) Band keratopathy.

b) Autoimmune endotheliopathy in inferior cornea.

c) Optic disc neovascularization.

d) Yellow, solid sub-retinal pigment epithelium detachments in the posterior pole.

6. Which one is not a characteristic finding in idiopathic intermediate uveitis?

a) Pars plana exudates.

b) Collagen band (snowbank) over the inferior pars plana.

c) Peripheral retinal vasculitis (periphlebitis/phlebitis).

d) Multiple, punched-out, pigmented chorioretinal lesions.

7. Which one of the systemic diseases has been associated with intermediate uveitis?

a) Multiple sclerosis.

b) Sarcoidosis.

c) Lyme disease.

d) All of the above.

8. Which laboratory investigation is most useful in intermediate uveitis?

a) ERG

b) HLA type testing.

c) Serologic testing.

d) Fluorescein angiography.

9. Which one is not a common therapeutic modality in the algorithm of treatment of intermediate uveitis?

a) Periocular and/or systemic corticosteroids.

b) Cryotherapy.

c) Intense topical corticosteroid therapy.

d) Immunosuppressive drugs.

e) Pars plana vitrectomy.

10. Which is not true about the snowbank in intermediate uveitis?

a) Histologically it consists of a collapsed, condensed vitreous, scattered lymphocytes, blood vessels, spindle-shaped cells, and hyperplastic nonpigmented ciliary body epithelium.

b) It is usually found over the inferior pars plana but can extend 360 degrees over the entire pars plana.

c) It is associated with a worse prognosis.

d) It is a sine qua non for the diagnosis of intermediate uveitis.

Answers

1.b 2.d 3.d 4.d 5.d 6.d 7.d 8.d 9.c 10.d