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A case report of multifocal choroiditis panuveitis

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Abstract : MCP is a condition characterized by intraocular inflammation and multifocal choroidal lesions occurring in the absence of any known ocular disease. MCP is one of the most common white dot chorioretinal inflammatory syndromes. The most common ocular manifestation of tuberculous infection is choroiditis. The cause of MCP is not fully understood. It is possible that an exogenous pathogen(s) whether viral, bacterial or fungal, initially triggers an immune response which can lead to subsequent exacerbations in the absence of the inciting pathogen. Here we present a case of Multifocal Tuberculous Choroiditis - Panuveitis in a young male.

Keyword : Multifocal Choroiditis, MCP, Choroiditis, Panuveitis, Tuberculosis, Pseudo POHS

Introduction:

MCP is a condition characterized by intraocular inflammation and multifocal choroidal lesions occurring in the absence of any known ocular disease. In 1973, Nozik and Dorsch described 2 patients with bilateral anterior uvetis associated with a chorioretinopathy resembling the presumed ocular histoplasmosis syndrome (POHS). In 1984, Dreyer and Gass presented their series of 28 patients with uveitis and similar lesions at the level of the retinal pigment epithelium (RPE) and choriocapillaris, and called the syndrome multifocal choroiditis and panuveitis. In 1985, Deutsch and Tessler described 28 patients with a similar condition, which they called inflammatory pseudohistoplasmosis, but most of these patients had findings to suggest systemic diseases such as sarcoidosis, syphilis, or tuberculosis. Finally, in 1986, Morgan and Schatz reported 11 similar cases of a condition they called recurrent multifocal choroiditis. These four reports all describe a condition with fundus lesions that mimics POHS but with the addition of vitritis and, often, an anterior uveitis. MCP is one of the most common white dot chorioretinal inflammatory syndromes. The most common ocular manifestation of tuberculous infection is choroiditis. Bouza et al, in Madrid, Spain reported that up to 17% of their patients with proven TB infection had signs of choroiditis, most of whom were asymptomatic. This concurs with the observation by Biswas et al in Madras, India that in a series of 1005 patients with proven systemic TB, only 1.4% developed ocular morbidity. Multifocal choroiditis was again the most common finding.

Epidemiology:

The disease is typically bilateral and seems to have a predilection for females in the second to sixth decades of life, with a median age of 28 to 33 years. Most patients give no history of living in areas endemic for POHS nor do they have affected family members. Most patients have bilateral involvement (45%-79%) but there may be asymmetric involvement and many of the involved second eyes may be completely asymptomatic. Patients usually present subacutely with decreased central vision or metamorphpsia. Other less common presenting complaints include paracentralscotomata, floaters, photopsias, mild ocular discomfort, and photophobia. Initial visual acuity is highly variable, ranging from 20/20 to light perception.

Clinical Profile:

Chief ailments:

14 years male, presented with complaints defective vision both eyes for 5 months

Presenting complaints:

Complaints of defective vision OU for 5 months – painless, progressive loss of vision both for near and distant vision; No history of double vision, transient obscuration of vision, defective colour vision, headache, nausea, vomiting, altered size perception of objects, flashes of light, floaters, defective dark adaptation, painful eye movements, loss of field of vision, brow ache, watering, irritation, photophobia, redness, colored halos, trauma

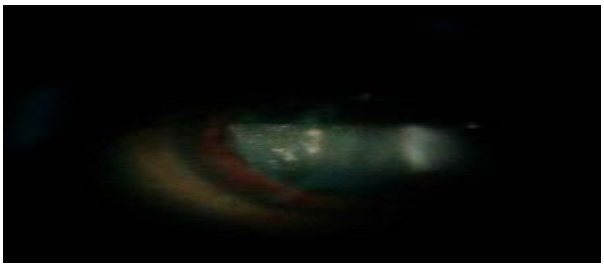
- H/o evening rise of temperature +
- H/o loss of weight; loss of appetite
- H/o night sweats +

Past History:

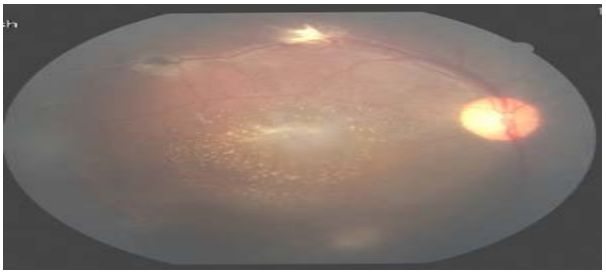
- Not a known diabetic, hypertensive, seizure disorder,
- Not on any prolonged topical/systemic medications
- No h/o any previous surgeries

Ocular Examination: OU
Lids – normal
Conjunctiva – Normal
Cornea – Mutton Fat KPs +
Anterior Chamber – Occasional cells +; Flare 2+
Iris – No evidence of iris nodules;
Pupils – 3 mm; round; reacting to both direct and consensual light reflex; No RAPD; Convergence +
Lens – Clear
Extra ocular movements – Full
Anterior Vitreous – Cells ++; Flare ++
UCVA – 6/12; BCVA – 6/6 p
IOP (by NCT) – 14 mm Hg
Central fields – Normal, no enlargement of blind spot; Colour Vision – Normal; no colour desaturation
Dilated Fundus Examination revealed: mild hazy media due to vitritis, with disc being vertically oval; normal in size; pink in colour; with well- defined margins maintaining a cup to disc ratio of 0.3 with a healthy neuro retinal rim. Several yellow lesions are seen at the level of the RPE and choriocapillaris seen in all quadrants with radiating linear streaks with areas of old chorio-retinal scarring in and around macula with absent foveal reflex suggestive of Multifocal Choroiditis – Panuveitis. Vitritis

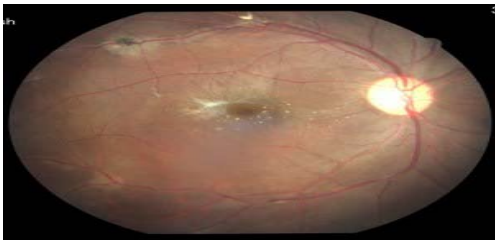
Vitritis OD



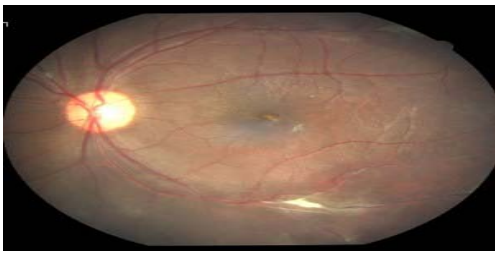
Vitritis OS



Pre Treatment Fundus - OD
Investigations:
Hemoglobin: 12.4 grams;
Complete Haemogram – Normal;
Renal Function – Normal;
ESR – elevated;
Mantoux Test – positive;
Chest X ray – PA view: suggestive of Pulmonary Tuberculosis;
Sputum AFB - Negative
Treatment:
Anti TB regimen – Category II (as per RNTCP guidelines) initiated; strict adherence to regimen emphasized;
Oral corticosteroids:
Initial dose: Tab. Prednisolone 1 mg / kg body weight started and tapered in weekly titrated doses.
Maximum dose: 60 – 80 mg/day
Maintenance dose: 10 mg/day
Tapering schedule: over 40 mg/day; decrease by 10 mg/ day every 1 – 2 weeks; 40 – 20 mg/day; decrease by 5 mg/day every 1 – 2 weeks; 20 – 10 mg/day; decrease by 2.5 mg/day every 1 – 2 weeks; 10 – 0 mg/day; decrease by 1 – 2.5 mg/day every 1 – 4 weeks. T. Calcium 1500 mg daily and Vit D 800 IU daily.
Follow up:
The patient was followed up subsequently after 2 weeks. Slit Lamp and Dilated fundus examination - OU revealed: Resolving vitritis with regression of fundus lesions with areas of chorio retinal scarring. Hence ATT was continued and drug compliance ensured. Corticosteroids tapered.



Fundus - Post Treatment - OD



Fundus - Post Treatment - OS
Discussion:
Etiology
The cause of MCP is not fully understood. Tiedeman suggested a viral etiology when he found serologic evidence of chronic or persistent Epstein-Barr virus (EBV) infection in 10 patients with MCP which he did not find in 8 control patients. A subsequent study by Spaide et al. did not support this hypothesis. Moreover, patients with MCP do not have systemic signs of chronic EBV infection. In another recent study 7 cases of MCP were evaluated. The presence of specific antibodies in the aqueous and serum suggested recent infection with herpes zoster in 2 cases and herpes simplex in 2 cases. Biswas et al in Madras, India that in a series of 1005 patients with proven systemic TB, with Multifocal choroiditisbeing the most common finding. It is possible that an exogenous pathogen(s) whether viral, bacterial or fungal, initially triggers an immune response which can lead to subsequent exacerbations in the absence of the inciting pathogen. Additional work is needed to more clearly define the cause of MCP.

Clinical Spectrum of the Disease:

Tuberculosis (TB) once a significant disease with dwindling incidence, is now again on the increase. Possible causative factors include a growing immunocompromised population, the emergence ofdrug-resistant strains, mass migration from endemic regions and the cessation of immunization programmes in some areas. Anti-TB drugs such as ethambutol and rifampicin occasionally cause visual deficit, thus an ophthalmological referral is helpful. In addition, TB may directly involve ocular structures. The uveal tract with its rich blood supply, is particularly susceptible to both metastatic infection and diseases with a predominantly immunological basis. *Anterior uveitis:* Anterior uveitis in MCP may consist of mild to moderate cellular reaction in the anterior chamber, granulomatous keratic precipitates, and posterior synechia. In POHS, the anterior chamber is notably clear. *Vitritis*: MCP differs from POHS clinically in a number of ways , the most important being the presence of vitritis in one or both eyes. In fact, the presence of vitreous cells in an eye effectively eliminates the diagnosis of POHS. Vitritis was observed in all of the patients in the series of Nozik and Dorsch, and Dreyer and Gass and in 89% of the patients in Deutsch and Tessler's series. The vitritis ranges from mild to moderate and may be asymmetrical. Little vitreous debris is seen once the active inflammation quiets down. *Chorioretinal lesions*: The key finding in MCP is the chorioretinal lesions scattered in the fundus. Acutely, several to several hundred yellow (sometimes gray) lesions are seen at the level of the RPE and choriocapillaris . Most lesions are 50 to 350 mm in diameter but occasionally may be larger. In contrast, the lesions in POHS tend to be larger (300 to 1000 mm) and fewer (less than 10).

The lesions are usually round or oval in shape and may be seen in the posterior , mid-peripheral or peripheral retina. They may be seen singly, in clusters, or arranged in a linear configuration (linear streaks) in the equatorial region . These lesions eventually become deep, round and atrophic, with variable degrees of pigmentation and scarring . New chorioretinal lesions may be seen in conjunction with old scars in recurrences. Typically, most patients develop peripapillary scarring similar to that seen in POHS (white, napkin-ring like). Optic disc pallor and narrowing of the retinal vessels is much less frequently seen. Cystoid macular edema (CME) occurs in 10% to 20% of patients. About 25% to 39% of patients with MCP develop macular and peripapillarychoroidal neovascular membranes (CNVM's) which may be the presenting cause for decreased vision. Macular CNVM with extensive scarring is the major cause of visual loss in MCP. In fluorescein angiography acute lesions hyperfluoresce early and leak lately. Old scars typically act as window defects with early hyperfluorescence and late fading. Visual field testing may reveal blind spot enlargement, scotomata corresponding to the chorioretinal lesions, or large temporal field defects which do not necessarily have corresponding fundus lesions.

Differential Diagnosis:

Presumed Ocular Histoplasmosis Syndrome (POHS): Examination during the active stage reveals inflammation in the anterior chamber and vitreous, an important finding distinguishing MCP from the POHS. The choroidal lesions may range from approximately 50 to 350 m in size and may be located in the posterior or peripheral fundus, concentrated in the macula or forming peripheral linear streaks. Active lesions appear as yellowish -white choroidal infiltrates and may be associated with neurosensory retinal detachments, vascular sheathing, disc edema, and cystoid macular edema. Inactive lesions appear as variably pigmented, punched-out areas of chorioretinal atrophy. Peripapillary scarring and extensive subretinal fibrosis are also common findings On fluorescein angiography active lesions may show early hypofluorescence with late hyperfluorescence. Cystoid macular edema is seen in 14% to 41% of affected eyes. ICG angiography shows more hypofluorescent lesions than observed by ophthalmoscopy. Visual field testing may show an enlarged blind spot and defects corresponding with the extent of multifocal lesions. Even in the absence of active disease the ERG may show progressive deterioration.

Ocular Sarcoidosis: The sole presence of multifocal choroiditis is usually suggestive of TB or sarcoidosis. TB choroiditis may involve any or all quadrants of the fundus while sarcoidchoroiditis tends to involve its inferior aspect. The typical radiological features of sarcoidosis include widespread pulmonary changes. Sarcoidosis however, is rare without hilar involvement and hence radiological evidence is essential.

Multiple evanescent white dot syndrome (MEWDS) primarily affects young women but the presentation is more often acutely, it is usually unilateral, the small gray-white lesions are at RPE level and usually confined to the posterior retina, there is typical orangish macular granularity. It is self-limited (average 8 weeks) with a return of visual acuity to 6/6 to 6/9, and it rarely recurs.

Punctuate inner choroidopathy (PIC), described by Watzke et al., resembles closely MCP. Patients with PIC are young, moderately myopic women and usually present acutely with blurred central vision, flashing lights and small central or paracentral scotomas. There is usually no intraocular inflammation, chorioretinal lesions appear all at one time, are confined to the posterior and midperipheral retina, and it does not usually recur, unlike MCP. Visual prognosis is good and no treatment is needed except for the 25% of eyes that develop a CNVM. Other diseases that should be considered in the differential diagnosis of MCP include syphilis, Lyme disease, intraocular lymphoma, herpes virus infection, inflammatory bowel disease and outer retinal toxoplasmosis. These diseases usually have other characteristic clinical and/or laboratory findings that help in differentiation.

Treatment:

This is an interesting case because regression of choroiditis follows commencement of anti-TB medication. In addition, the lack of intraocular inflammation suggests that the choroiditis is probably infective rather than immunogenic in origin. The routeto infection is presumed to be haematogenous from the lung. The temporal sequence from the initial pulmonary involvement by TB to the choroiditis is probably due to the initial under-treatment of his pulmonary TB and his increased immunocompromisedstate. As the natural history of TB choroiditis is one in which visual morbidity may not occur, anti-TB medication is initiated when systemic steroid is required to control significant intraocular inflammation or when there is a significant possibility that the patient has miliary or recurrence of systemic TB. The prognosis for MCP is guarded. The active disease course may be short, with few lesions, or chronic, with recurrent exacerbations of intraocular inflammation and the development of numerous lesions, progressive scarring, and retinal degeneration. Cystoid macular edema, subretinal fibrosis, and choroidal neovascularization are common causes of visual loss. Treatment relies on the use of corticosteroids (oral or periocular) with the use of other immunosuppressive agents (cyclosporine, azathioprine, methotrexate, chlorambucil, cyclophosphamide) as needed depending on the severity of the disease. Management of choroidal neovascularization, a poor prognostic sign, can be difficult as the best treatment is yet to be determined and spontaneous regression may occur. The current treatment options include periocular, intraocular, or oral corticosteroids; laser photocoagulation; photodynamic therapy; and, most recently, anti-VEGF therapy.

References:

1. Dreyer RF and JDM Gass. Multifocal choroiditits and panuveitis: a syndrome that mimics ocular histoplasmosis. Archives of Ophthalmology 1984;102:1776-1784.

2. Deutch TA and HH Tessler. Inflammatory pseudohistoplasmosis. Annals of Ophthalmology 1985;17:461-465.

3. Watzke RC, Packer AJ, Folk JC, Benson WE, Burgess D, and RR Ober. Punctate inner choroidopathy. Am J Ophthalmology 1984;98:572-584.

4. Vandenbelt, S, Shaikh, S, Capone A, and G Williams. Multifocal choroiditis associated with west nile virus encephalitis. Retina 2003;23:97-99.

5. Goldstein DA, Fontanilla FA, Kaul S, Sahin O, and HH Tessler. Long-term follow-up of patients treated with short-term high-dose chlorambucil for sight-threatening ocular inflammation. Ophthalmology 2003;109(9):370-377.

6. Samson CM, Waheed N, Baltatzis S, and CS Foster. Methotrexate therapy for chronic non-infectious uveitis: analysis of a case series of 160 patients. Ophthalmology 2001; 108(6): 1134-1139.

7. Kaplan-Messas A, Barkana Y, Avni I, and R. Neumann. Methotrexate as a first-line corticosteroidssparing therapy in a cohort of uveitis and scleritis. Ocular Immunology and Inflammation 2003; 11(2): 131-139.

8. Spaide RF, Freund KB, Slakter J, Sorenson J, Yannuzzi LA and Y Fisher. Treatment of subfoveal choroidal neovascularization associated with multifocal choroiditis and panuveitis with photodynamic therapy. Retina 2003;22(5):545-549.

9. Wachtlin J, Heimann H, Behme T, and MH Forester. Long Term results after photodynamic therapy with verteporfin for choroidal neovascularizations secondary to inflammatory chorioretinal diseases. Graefe's Arch Clin Exp Ophthalmol 2003;241(11): 899-906.

10. Brown, J., Folk, JC, Reddy, CV, and AE Kimura. Visual prognosis of multifocal choroiditis, punctate inner choriodopathy, and diffuse subretinal fibrosis syndrome. Ophthalmology 1996;103: 1100-1105.