Macular CMV Retinitis

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Introduction

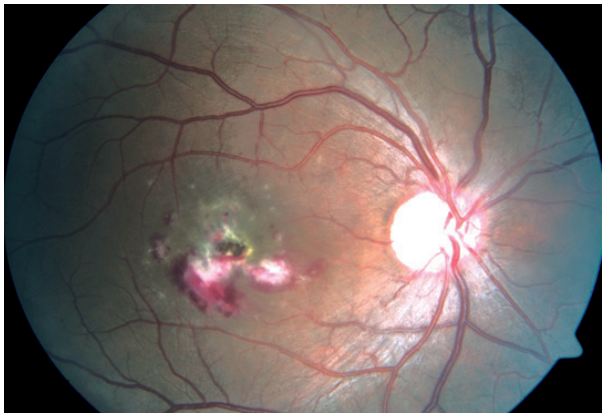
Acquired Immunodeficiency Syndrome is a potentially fatal multisystem syndrome, characterized by profound disruption of the immune system and a propensity for various opportunistic infections and neoplasms. Ocular involvement occurs in up to 73% of AIDS patients [1, 2] with the most common lesions being a retinal vasculopathy consisting of cotton-wool spots, retinal hemorrhages, and infectious retinopathy such as cytomegalovirus (CMV), herpetic, toxoplasmic or luetic retinitis.

CMV infection is a major cause of morbidity and mortality in AIDS. CMV retinitis has been reported to occur in 15-40% of AIDS patients with the rate declining since the arrival of HAART[3, 4,5]. CMV is a slowly progressive necrotizing retinitis that may affect the posterior pole, the periphery, or both, and, may be unilateral or bilateral. CMV retinitis more commonly involves retinal periphery. Here we present a rare case of CMV retinitis with isolated macular involvement.

Case Report

A 38 year old male patient presented to us with complaints of sudden onset of painless defective vision in his right eye for past 15 days. He was a retro positive patient on HAART therapy for the past 8yrs. Patient gave history of taking antituberculous therapy in 2007. He was diagnosed with immunological failure and has started on second line antiretroviral therapy as his CD4 count was low (114) and viral load was high (> 4lakh). He gave no history of any other systemic illness.

On ocular examination, his right eye showed a BCVA of 6/60, with 1+ cells in the anterior vitreous face. Rest of the anterior segment

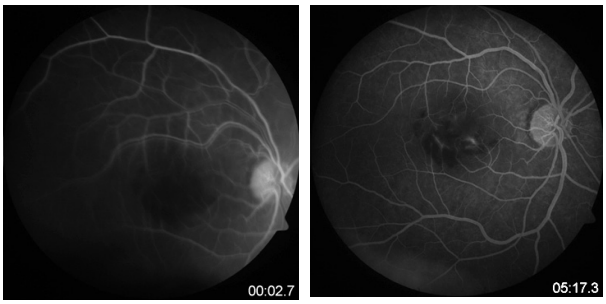


RE

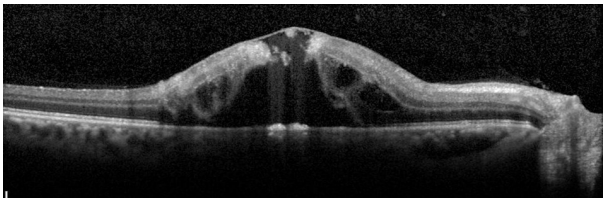
examination of the right eye was within normal limits. Anterior segment examination of the LE was within normal limits with a BCVA of 6/6. Posterior segment examination of the RE showed. A small central scar at macula surrounded by white retinitis patches and retinal haemorrhages along with few satellite lesions, clinically suggestive of CMV retinitis.

Posterior segment examination of the left eye was within normal limits.

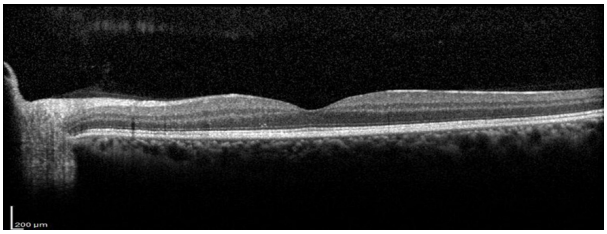
Blood investigations were done which showed, an elevated ESR (85mm/hr) and CRP (20.7mg %) and slightly reduced RBC counts (3.81million/µl). VDRL and mantoux were negative. FFA and OCT were done which showed.



FFA of RE shows blocked haemorrhage at macula due to retinal haemorrhage in early phase and minimal staining due to scar in late phase.



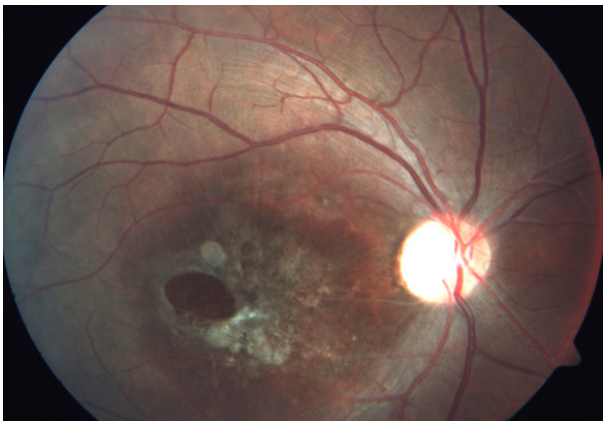
OCT RE: Thinned out retina at macula due to necrosis with ILM drape.



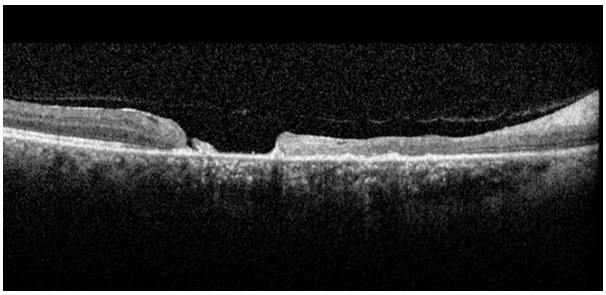
LE OCT: showed normal foveal contour.

Right eye vitreous tap was done and PCR was sent for HSV, CMV, VZV, TOXO, M.TB .The patient was started on T.Valcivir 1gm tid and was advised to continue ART and review with reports after one week. PCR was negative for HSV, CMV, VZV, TOXO, M.TB. The patient was advised to continue T.Valcivir and ART. Subsequent follow up visits showed signs of resolving lesion. CD4 count (last follow up) improved from 114 to 612.

On the last follow up, anterior segment examination of RE showed a BCVA of 6/24 (improved from 6/60 at presentation) and a quiet AVF. LE was within normal limits. Posterior segment examination of RE showed full thickness macular hole with scar at the macula along with epiretinal membrane.



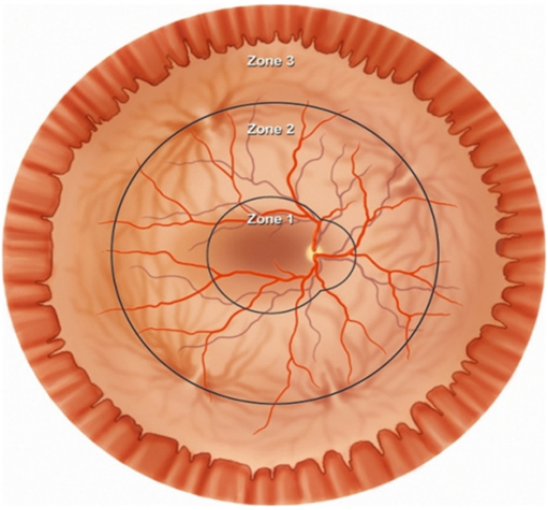
RE



Discussion

The prevalence of CMV retinitis is more in AIDS than in other immunosuppressed conditions. A prospective study conducted in India revealed that 17% of the patients with AIDS had CMV retinitis. HAART leads to reduced HIV replication, increase in CD4+ T-lymphocyte counts, reduced incidence of opportunistic infections such as CMV retinitis. Since the use of HAART, the incidence of CMV retinitis has declined by 50% to 80%. Risk for developing CMV retinitis is increased when CD4+ counts < 50/mm3 (by approx. 20%/year). Active CMVR in patients with higher CD4 counts can occur through destruction of CMV-specific CD4 memory cells.

HIV microvasculopathy with cotton wool spots has been demonstrated as an independent



Zones In CMV Retinitis
Zone 1: area within one DD of ONH or 2 DD of fovea. Zone 1 lesions are sight threatening and require immediate attention.

risk factor for CMV retinitis. It typically presents with peripheral areas of retinal whitening, often associated with retinal hemorrhages, which then progresses in a "brushfire-like" extension across the posterior pole.

CMVR is usually diagnosed clinically. In general, laboratory tests, such as CMV serology, antigen detection, PCR, or viral blood cultures, do not play a role in the acute evaluation in this setting. In routine cases, it is not necessary to obtain a retinal biopsy or attempt to identify CMV in the eye to make the diagnosis. In unusual cases, when diagnosis is not clear or when the patient does not respond to initial therapy, identifying CMV DNA in vitreous or aqueous humor, or rarely in tissue obtained via endoretinal biopsy, may help to establish the diagnosis.

The management of CMV retinitis is based on the recommendations from the CDC, the National Institutes of Health, and the HIV Medicine Association/Infectious Diseases Society of America, which state that "the choice of initial therapy for CMV retinitis should be individualized based on the location and severity of the lesion(s), the level of underlying immune suppression, and other factors such as concomitant medications and ability to adhere to treatment".

A. In patients with immune recovery (CD4+ count > 50 cells/mL or > 100 cells/mL for at least 3–6 months): discontinuation of maintenance therapy for inactive CMV retinitis is usually possible to avoid cost and toxicity of treatment.

B. In patients with immediate site-threatening retinitis: oral valganciclovir + intraocular ganciclovir implant for initial therapy (or) oral valganciclovir alone for initial therapy.

C. Patients who do not have site-threatening lesions: oral valganciclovir alone as initial therapy, in view of ease of administration and lack of requirement for ophthalmologic surgery or placement of an indwelling intravenous catheter.

Conclusion

CMV retinitis in almost all cases is a blinding disease if not controlled. It typically presents in the retinal periphery. Macula is rarely affected first in CMV retinitis and isolated macular lesions are rare, which highlights the rarity of this case showing an isolated Macular CMV retinitis. Thus a diagnosis of CMV retinitis has to be taken into account in all macular confined lesions in immunosuppressed patients. An early and accurate recognition is imperative to avoid destruction of the posterior pole and optic nerve head which can lead to complete blindness.

References

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