CASE REPORT

Optic Neuritis and Retinal Vasculitis as Primary Manifestations of Systemic Lupus Erythematosus

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Summary

Systemic lupus erythematosus (SLE) is a common multisystem disorder. However, retinal vasculitis as a primary manifestation of SLE is uncommon, accounting for only 4% of causes of retinal vasculitis. The postulated mechanism appeared to be vaso-occlusion of the retinal arterioles by thrombosis, with resultant ischaemia. Optic neuropathy in SLE is also rare, with a prevalence of 1%. This is a case report of a young lady who presented to us with retinal vasculitis as her initial presentation of SLE. Interestingly, the pathologic mechanism appeared to be inflammatory and not vaso-occlusive.

Case Report

A 19-year-old Malay lady presented with sudden, painless loss of vision in the left eye, particularly affecting the central field of 4 days' duration. It was associated with floaters. Systemic review revealed that she has been having recurrent mouth ulcer, hair loss and arthralgia for the past 2 years.

On examination of the left eye, visual acuity was counting fingers. Relative afferent pupillary defect was elicited. Anterior chamber examination did not show any activity. Fundus examination revealed mild vitritis, with hyperaemic and swollen optic disc. The retinal veins exhibited extensive periphlebitis and associated exudative macular detachment (Fig. 1). Visual field examination with Goldmann perimetry revealed central scotoma. Fluorescein angiography showed extensive perivascular leak. However, there were no areas of capillary non-perfusion. The right eye was essentially normal.

The initial blood investigations revealed an erythrocyte sedimentation rate of 95mm in first hour, a white blood count of 8.9g/L with a normal differential count and normal renal profile. Chest x-ray was normal. Tests for sarcoidosis were negative. The patient was treated with intravenous methylprednisolone 250mg 6-hourly for 3 days, followed by oral prednisolone of 1 mg/kg/day (60 mg/daily) for one week, which was then gradually tapered. Further diagnostic evaluation in this patient showed a positive antinuclear antibody value and a double stranded DNA, diagnostic of SLE.

This article was accepted: 8 July 2002

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Based on the clinical findings and serology results, a diagnosis of left retinal periphlebitis and exudative macular detachment with optic neuritis secondary to SLE was made. The patient improved clinically with corticosteroid therapy, with gradual tapering done over 3 months (Fig. 2). She was referred to the physician for further management of her SLE. Her best-corrected vision in the left eye was 6/18 and N5 at 8 months later.

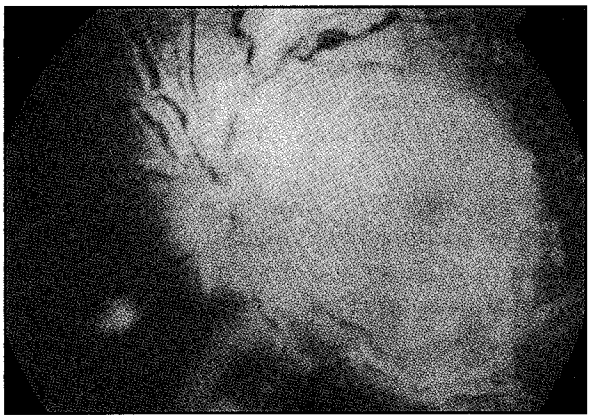

Fig. 1: Left fundus at presentation showing mild vitritis, a swollen and hyperaemic disc, extensive periphlebitis, and exudative macular detachment

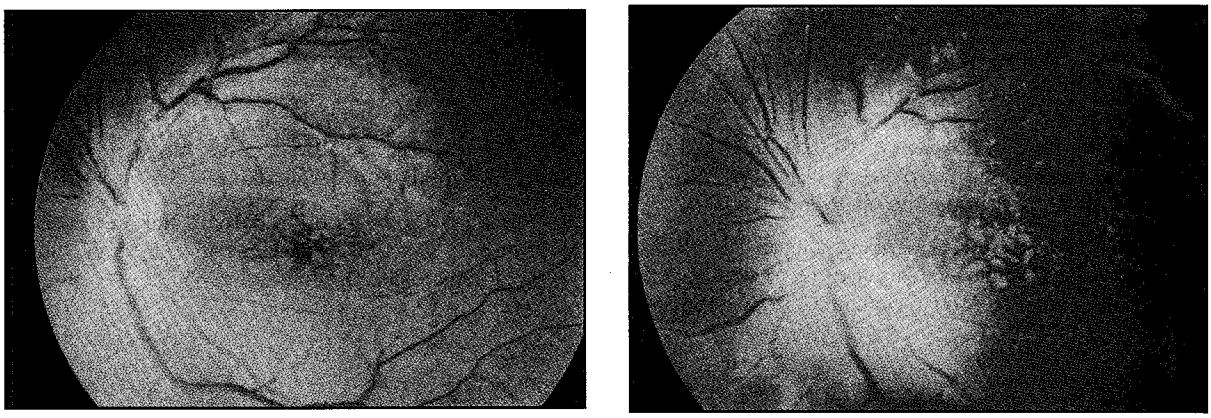

Fig. 2: Left fundus appearance at 1 month and 3 months showing resolution of periphlebitis and exudative detachment over the macula

Discussion

Retinal manifestations of SLE are well-documented complications of SLE. The prevalence in the western literature varies between 5-16%[1,2]. It is dominated by a microangiopathy, thought to be of an immune complex-mediated vascular insult, with occlusive consequences. In its mildest form, which is the commonest, it consists of cotton wool spots with/without haemorrhages due to focal retinal ischaemia. More severe retinal vaso-occlusive disease, termed retinal vasculitis affects the main arterioles[2]. Patients present with central retinal artery occlusion or vein occlusion with resultant severe visual loss, often associated with central

nervous system lupus. Proliferative lupus retinopathy,[3] that is rare, is characterized by disc and retinal neovascularization occurring in response to severe retinal ischaemia.

The term lupus vasculitis is commonly used in clinical practice: however, in the Graham study [4] it was shown that the underlying pathologic abnormality of the retinal arterioles is that of arteriolar occlusion by fibrinoid material and not inflammation.

Our patient presented with severe periphlebitis, with resultant exudative macular detachment and concomitant optic neuritis. Retinal haemorrhages and cotton-wool spots, typical of SLE were not observed. There were no signs of retinal ischaemia, clinically, as well as angiographically. This perhaps represent the first documented case of retinal periphlebitis attributed to SLE, with the underlying responsible pathogenic mechanism not of vaso-occlusive in nature, but rather truly vasculitic in nature.

This case illustrates to us, the ophthalmologists, that retinopathy can be a primary presenting manifestation in an SLE patient who is otherwise healthy. Bearing this in mind, apparently healthy young patients presenting with retinal vasculitis should be worked up for SLE..

References

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