

# Recalcitrant Rubeosis Iridis in Systemic Lupus Erythematosus

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*Unilateral ischaemic retinopathy resulting in vitreous haemorrhage and neovascular glaucoma is an uncommon presentation in systemic lupus erythematosus. This report describes a patient with systemic lupus erythematosus with recalcitrant rubeosis iridis.*

**Key words:** Lupus erythematosus, systemic, Neovascular glaucoma, Retinal diseases, Vitreous hemorrhage

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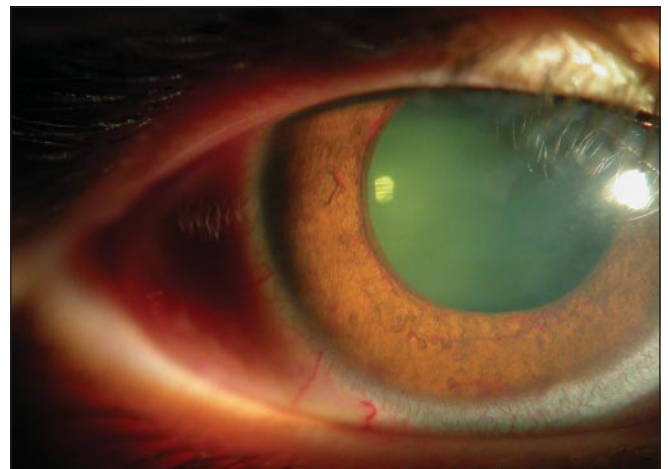
## Introduction

We describe a patient with systemic lupus erythematosus (SLE) with recalcitrant unilateral retinal ischaemia resulting in proliferative changes complicated by vitreous haemorrhage and neovascular glaucoma.

## Case Report

This report describes a 17-year-old Chinese boy with both lupus nephritis and cerebral lupus. The diagnosis of SLE was based on the American College of Rheumatology criteria. The patient first presented to the Department of Ophthalmology, Penang Hospital, Penang, Malaysia, in April 2005 with vision of counting fingers at 1 metre without obvious relative apparent pupillary defect in the right eye. Fundoscopy revealed extensive cotton-wool spots with a normal looking macula and optic disc. The left eye was normal. Fundus fluorescein angiography was not performed because of associated renal impairment. Pan-retinal photocoagulation (PRP) was performed. In August, new vessels were noted in the disc and repeat PRP was performed (Figure 1). Although his erythrocyte sedimentation rate (ESR) was normal and thrombotic screen was negative, he was given oral prednisolone 1 mg/kg daily and aspirin 150 mg daily. After 1 month, he developed neovascular glaucoma and vitreous haemorrhage. He underwent pars plana vitrectomy and endolaser. Unfortunately, the rubeosis worsened and he required intraocular pressure control with diode cycloablation. In December, the vitreous haemorrhage recurred and the eye became phthisical.

Figure 1. Rubeosis iridis developing 1 month after new vessels at the disc were noted.



## Discussion

Retinal ischaemia characterised by cotton-wool spots with or without intraretinal haemorrhages is not an uncommon manifestation of SLE, but severe ischaemia resulting in proliferative retinopathy and blindness is less common.<sup>1</sup> Unilateral disease is rare. This case report illustrates that unilateral rubeosis iridis can occur in an ischaemic retina secondary to SLE. The presence of rubeosis heralds a poor visual outcome for the majority of patients.<sup>2</sup> Irreversible visual loss in these patients is typically due to macular ischaemia and complications of the proliferation such as vitreous haemorrhage and neovascular glaucoma.

The mechanism for retinal ischaemia is probably a combination of vaso-occlusion by thrombosis and inflammation; therefore, treatment with both an antithrombotic agent and an anti-inflammatory agent is required. This report also shows that the progression from symptomatic retinal ischaemia to uncontrolled neovascular

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glaucoma can occur within months, resulting in rapid progression to blindness.

Eighty six percent of retinopathies are associated with the presence of anticardiolipin antibodies, which cause arterial and venous thrombosis.<sup>3</sup> However, there was no evidence of thrombotic tendency or presence of anticardiolipin antibodies in this patient.

Patients with cerebral or central nervous system (CNS) involvement also tend to develop retinopathy, probably denoting a common pathogenic mechanism of disease such as the presence of anticardiolipin antibodies.<sup>3,4</sup> The CNS-retina association is seen in this patient, but again the antibodies were not detected.

This report also supports the evidence that retinal ischaemia in SLE is difficult to treat, even with combination medications and PRP. The ideal treatment for these patients has not yet been found.

Ocular manifestations are often said to be a reflection of systemic disease activity.<sup>1,3</sup> However, retinal ischaemia may be predominantly secondary to vaso-occlusion by thrombosis. Therefore, ESR, which is a sensitive test for identifying systemic activity, is not always the most appropriate marker for ocular disease activity if the inflammatory component is minimal.<sup>5</sup> A normal ESR is not always indicative of quiescent systemic disease. A more accurate means of determining disease activity would be to use the European Consensus Lupus Activity Measurement (ECLAM) score, which has 15 clinical and laboratory variables including ESR.<sup>6</sup> However, ECLAM scoring is time-consuming and costly for a busy government

hospital to perform. Systemic management can be tailored to the funduscopy findings rather than to the ESR level alone.

It may be prudent for the American College of Rheumatology to look into whether retinal manifestations may be used as one of the 12 criteria in diagnosing SLE, in view of the strong association of retinopathy with systemic disease activity.

This report illustrates the difficulty in treating severe retinal ischaemia in patients with SLE. The disease rapidly progresses to neovascular glaucoma and blindness. ESR alone is not an adequate marker of ocular disease activity.

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