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Case report

Retinopathy in lupus transitioned to Kikuchi-Fujimoto disease

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ABSTRACT

Purpose: We present a patient with systemic lupus erythematosus with significant vaso-occlusive retinal findings mimicking antiphospholipid antibody syndrome, who developed Kikuchi-Fujimoto disease.
Observations: Our patient was initially diagnosed with systemic lupus erythematosus with antiphospholipid antibody syndrome given consistent serologic markers and profound retinal vascular ischemia. However, on subsequent follow up, she presented with fever and lymphadenopathy and underwent lymph node biopsy, which declared histologic findings of Kikuchi-Fujimoto disease. Repeat markers for antiphospholipid antibody syndrome were negative and she was taken off lifelong anticoagulation.
Conclusions and importance: Systemic lupus erythematosus and Kikuchi-Fujimoto disease may have many similar features and even biomarkers, and given the potential overlap of presentation, clinicians must carefully distinguish between these diseases to prevent unnecessary treatment.

1. Introduction

Classically, Kikuchi-Fujimoto Disease (KFD) is characterized by fever and tender lymphadenopathy, typically self-limited over a period of a few months without treatment, often in young women of Asian heritage.[1,2] Systemic lupus erythematosus (SLE) and KFD have been reported to occur in patients at any time interval, including concurrently and sequentially to each other.[3–5] We present a case of a patient with SLE who presented with profound retinal ischemia mimicking antiphospholipid antibody syndrome (APLS), who developed KFD. Lymph node biopsy secured the diagnosis of KFD. Furthermore, the patient's initially elevated anticardiolipin antibodies when she presented with lupus flare were negative on repeat testing for APLS when she presented with clinical findings of KFD. This case did not require approval by our Institutional Review Board.

2. Case report

Specific personal identifying information has been removed from this report in accordance with the patient's wishes. A twenty-six-year old female, recently diagnosed and treated for SLE complicated by aseptic meningitis and multiple small strokes at an outside hospital 2 weeks prior, presented to our institution with complaints of painless decreased vision and right visual field deficit of the left eye (OS), occurring suddenly 12 days prior and without improvement in the intervening period. Visual acuity (VA) was 20/20 right eye (OD) and count fingers at 1 foot OS. Pupils were equal, round and reactive with a positive afferent pupillary defect OS. Brightness sense was decreased OS compared to OD and she was unable to perform color plates OS compared to 8/8 plates OD. Intraocular pressures as well as anterior segment exam were normal in both eyes (OU). Dilated fundus exam was significant for mild disc edema and a cotton wool spot along the superior arcade OD (Fig. 1). There was severe disc edema with peripapillary hemorrhage OS and an apparent cherry red spot in the macula. The vessels were sclerosed and sheathed, with many ghost vessels visible. There was diffuse retinal ischemia and retinal hemorrhages temporally in the periphery (Fig. 2). Optical coherence tomography (OCT) showed nerve fiber layer thickening corresponding to cotton wool spots. Fluorescein angiography (FA) demonstrated marked ischemia with complete nonperfusion extending from the macula (Fig. 3), in contrast to the normal perfusion in the right eye (Fig. 4). Work up revealed positive ANA (1:160, speckled), anti-smith (>8), anti-RNP (>8), SSA (3.1), anti-cardiolipin (26), with low C3 (39) and C4 (8.3) and she was diagnosed with SLE with APLS. Hypercoagulability workup including homocysteine, PT/INR, PTT were normal.

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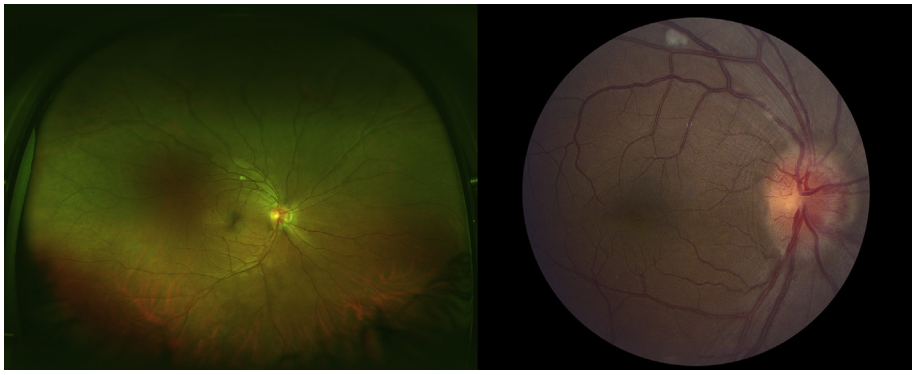

Fig. 1. Color fundus photo, right eye. Dilated fundus exam of the right eye was significant for mild disc edema and a cotton wool spot along the superior arcade.

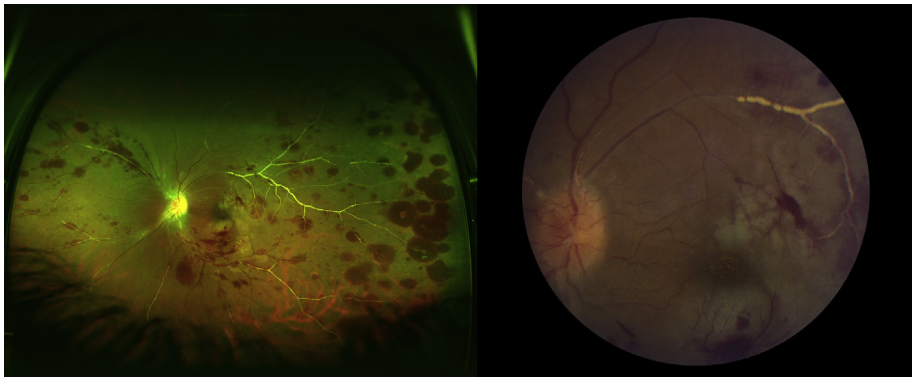

Fig. 2. Color fundus photo, left eye. Dilated fundus exam of the left eye was remarkable for severe disc edema with peripapillary hemorrhage, an apparent cherry red spot in the macula, sclerosed and sheathed vessels, diffuse retinal ischemia and retinal hemorrhages.

Further extensive infectious work up was negative for HIV, RPR, Quantiferon Gold, blood cultures, chest X-ray, lumbar puncture, and trans-esophageal echocardiogram. MRI was consistent with a punctate left basal ganglia lacunar infarct. MRA was normal. She was treated with high dose IV methylprednisolone starting at 1gm for a day and quickly tapered due to presumed steroid-induced psychosis to 500 mg for 2 days, then transitioned to prednisone 20 mg every 8 hours, and she was discharged on cyclophosphamide and hydroxychloroquine because of perceived severity of her SLE, and enoxaparin was transitioned to warfarin for life long

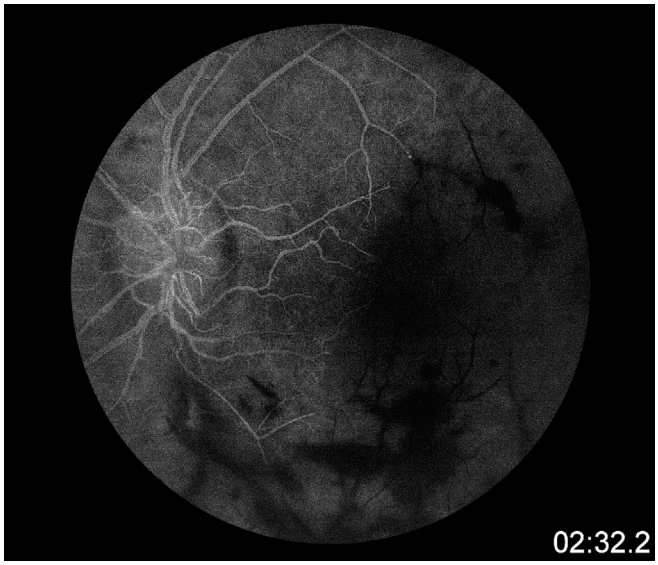

Fig. 3. Fluorescein angiography, left eye. Fluorescein angiography demonstrated marked ischemia with complete nonperfusion extending from the macula of the left eye.

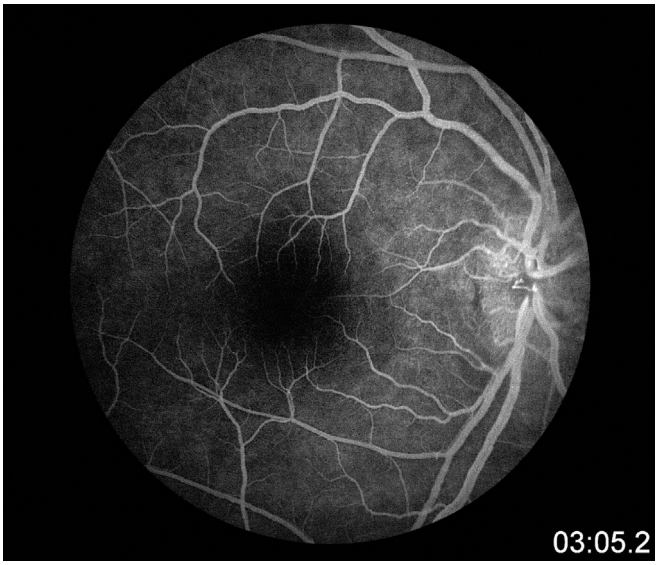

Fig. 4. Fluorescein angiography, right eye. Normal perfusion.