Purtscher-like retinopathy in a patient with lupus: a case report

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Case report

In January 2020, a 32-year-old Filipino woman was admitted to the medical unit of our hospital with subacute onset of a facial malar rash, digital vasculitis purpura and alopecia. Systemic lupus erythematosus (SLE) had been newly diagnosed according to the Systemic Lupus International Collaborating Clinics criteria. Initial blood tests revealed an elevated serum anti-ds DNA antibody level, positive anti-SM antibody, low C3 and C4 as well as pancytopenia. Fasting glucose and lipid profile were unremarkable. She was first treated with oral hydroxychloroquine 200 mg daily and oral prednisolone 20 mg daily. One week later, she presented to our ophthalmology clinic with acute-onset painless blurring of vision in her right eye upon waking that morning. Physical examination revealed a best-corrected visual acuity of finger counting at 30 cm in her right eye and 6/6 in her left eye with a right relative afferent pupillary defect. Slit-lamp examination was unremarkable with no anterior chamber inflammation. Fundus examination revealed numerous Purtscher flecken, which are polygonal patches of retinal whitening with distinct border and normal retina in between, across the posterior pole. A pseudo cherry-red spot and confluent retinal whitening could be seen in the right eye secondary to proximal occlusion of the retinal arteries. A few flame-shaped retinal haemorrhages were observed inferior and temporal to the optic disc in the right eye. The right optic disc was mildly pale with sharp margins and the retinal vessels were not tortuous, while the left optic disc is pink (Fig 1).

Urgent optical coherence tomography of the right eye showed hyper-reflectivity in the retinal nerve fibre layer and areas of retinal thickening. Fundus fluorescein angiography (FFA) of the patient's right eye showed capillary non-perfusion corresponding to Purtscher flecken, superior branch retinal arterial occlusion, and peripapillary staining (Fig 2). Optical coherence tomography and FFA of the patient's left eye were normal. Given the typical fundus appearance and FFA findings, our provisional diagnosis was Purtscher-like retinopathy with branch retinal artery occlusion related to SLE. We advised her medical team to escalate her systemic treatment immediately and she was given intravenous methylprednisolone 500 mg daily for 3 days, followed by oral cyclophosphamide 500 mg for 1 day before switching to oral prednisolone 40 mg daily. Oral hydroxychloroquine 200 mg daily was continued. On high-dose steroids, she developed features of psychosis that subsequently resolved. Magnetic resonance imaging of the brain showed no features typical of central nervous system lupus such as ischaemia and vasculitis. We closely monitored her condition and visual acuity had not improved 1 month after systemic treatment was started, remaining at finger counting at 30 cm. Fundus examination of the right eye after treatment revealed macula oedema and pseudo cherry-red spot, with no signs of optic atrophy. Cotton wool spots in the left eye had resolved. The patient chose to seek further medical attention overseas.

Discussion

At least one third of patients with SLE have ophthalmological involvement. Retinal and choroidal pathology are sinister ocular complications of SLE that can lead to permanently impaired visual acuity. Purtscher-like retinopathy is a type of vaso-occlusive retinopathy associated with multiple conditions including connective tissue disorders, pancreatic disease, renal disease, and haematological disease.[1]

We report this rare case of Purtscher-like retinopathy in a patient with SLE which had a devastating visual outcome despite systemic treatment. In an observational case series of 5688 patients with SLE, eight cases of Purtscher-like retinopathy were diagnosed.[1] All patients had received treatment but most had optic atrophy and persistent low visual acuity,[1] consistent with the outcome for our patient. Poor visual acuity has also been attributed to presentation with a pseudo

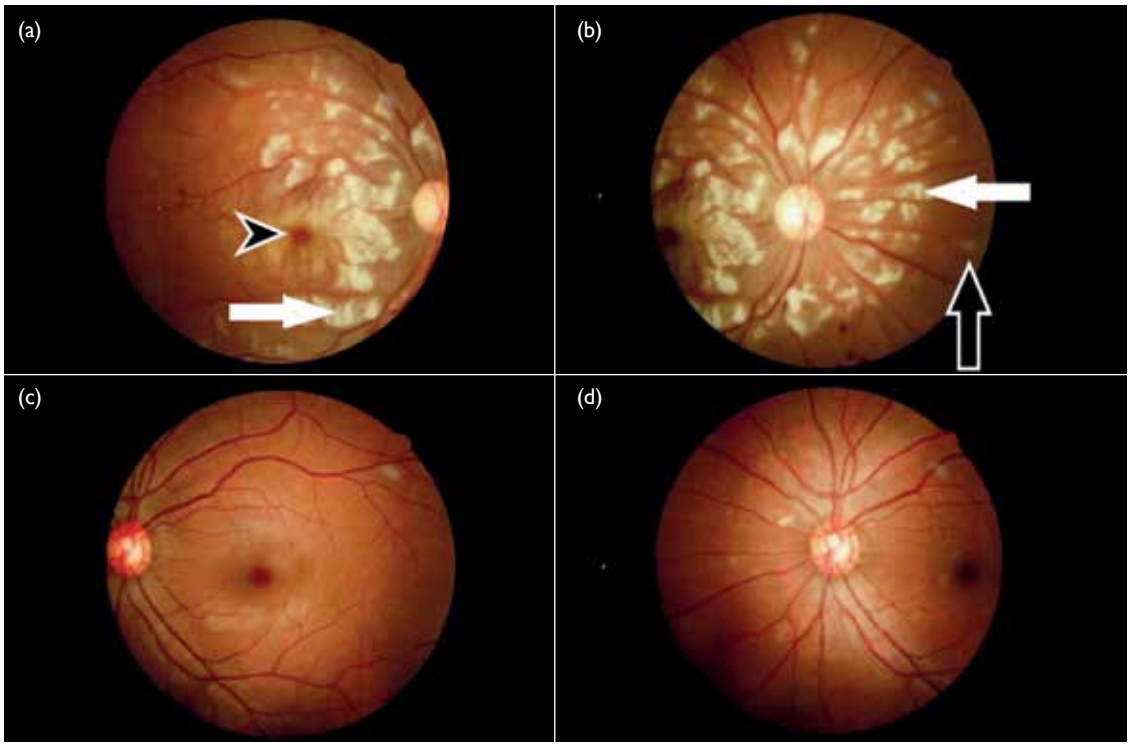

FIG 1. A 32-year-old woman with systemic lupus erythematosus. Fundus photographs of (a, b) the patient's right eye showing Purtscher flecken (closed arrows), cotton wool spots (open arrow), and a pseudo cherry-red spot (arrowhead); and (c, d) the patient's left eye showing no unusual findings

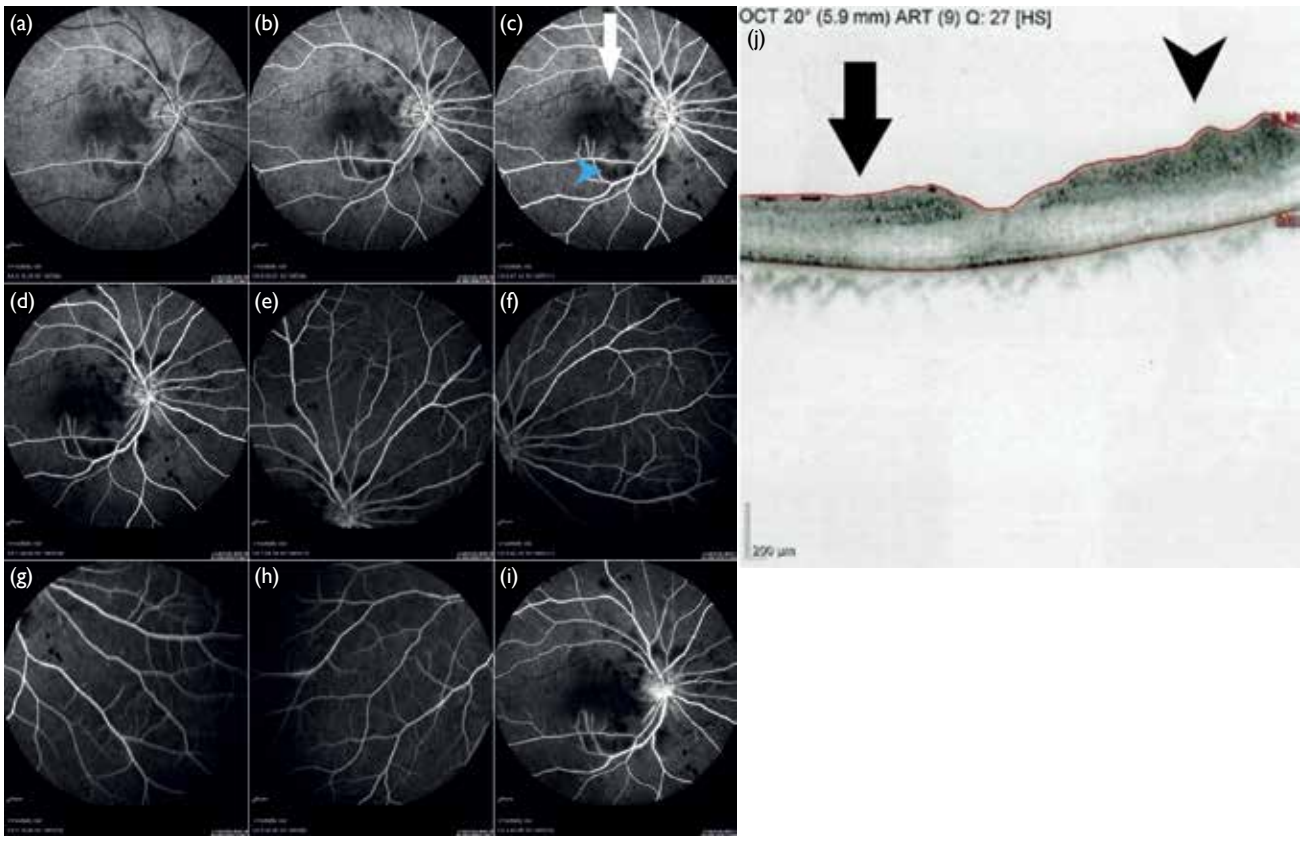

FIG 2. Same patient. (a-i) Fundus fluorescein angiography images of the patient's right eye showing superior branch retinal artery occlusion (white arrow) and areas of capillary non-perfusion (blue arrowhead). (j) optical coherence tomography of the patient's right eye showing hyper-reflectivity in the retinal nerve fibre layer (arrow) and areas of retinal thickening (arrowhead)

cherry-red spot. The prognosis of Purtscher-like retinopathy is better in patients with other underlying diseases. For example, Shahlaee et al[2] reported a case of postviral Purtscher-like retinopathy presenting with finger count visual acuity in both eyes. No treatment was given but the patient's final visual acuity improved to 20/20 with bilateral scotoma on visual field assessment. Another case of Purtscher-like retinopathy in a patient with adult-onset Still's Disease was reported by Yachoui,[3] in which initial visual acuity was 20/20 in the right eye and 20/25 in the left. Oral prednisolone 60 mg daily was initiated and the patient's final visual acuity was 20/25 in both eyes with evidence of established bilateral optic atrophy.

In a retrospective case control study, Gao et al[4] reported a 0.66% prevalence of retinal vasculopathy among SLE patients with signs ranging from cotton wool spots, retinal vascular attenuation to retinal haemorrhages. Lupus retinopathy can result in severe complications such as neovascularisation, macula oedema and retinal vessel occlusion resulting in vision loss.[4] It is known that patients with high Systemic Lupus Erythematosus Activity Disease Index score are at higher risk of Purtscher-like retinopathy, central nervous system lupus[1] and vaso-occlusive retinopathy, reflecting severe systemic microangiopathy.[1,4]

Because patients with active lupus are at risk of Purtscher-like retinopathy that can lead to severe irreversible visual loss,[1] it is important to raise awareness in patients with active lupus and visual symptoms. A high index of suspicion for retinopathy is needed for those with very active disease and prompt referral to an ophthalmologist is warranted in the presence of visual symptoms. Despite escalation of treatment after the onset of visual symptoms, our patient's vision did not recover. Early aggressive treatment to control SLE disease activity is essential to prevent this blinding complication.

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