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Purtscher-Like Retinopathy in a Children with Systemic Lupus Erythematous (SLE)

Abstract – Purtscher-like retinopathy is a rare ocular manifestation in systemic lupus erythematosus (SLE). **Objective:** To report a case of Purtscher-like retinopathy in a 10-year-old female child with SLE. **Case presentation:** A 10-year-old Chinese girl was presented with lethargy for four weeks and intermittent fever for two weeks. The fever was associated with multiple body rashes. She was treated as pyrexia of unknown origin. Generally she was febrile with temperature of 39°C, presence of facial rash and discoid rashes involving both hands with generalised erythematous skin over the hernial region. Patient was referred by the paediatrics team to rule out any ocular association of connective tissue disease. Fundus examination showed presence of generalised Purtscher-like retinopathy. Blood investigation revealed leukopenia, anaemia, low complement (C) 3 (C3) and C4 with positive both antinuclear antibody (ANA) and anti-double-stranded DNA (anti-dsDNA) antibody. She was finally diagnosed as childhood-onset systemic lupus erythematous (SLE) and was treated systemically with prednisolone, mycophenolate mofetil (MMF) and hydrochloroquine by paediatric team. There was improvement of visual acuity with resolution of Purtscher-like retinopathy at one year review. **Conclusion:** Childhood-onset SLE presented without visual complaints must be evaluated thoroughly for Purtscher-like retinopathy. Early recognition and treatment lead to a better visual outcome.

Keywords – *Purtscher-like retinopathy, systemic lupus erythematous*

1 INTRODUCTION

Purtscher retinopathy was first described by Otmar Purtscher in patients with rapid sight loss following a severe head injury (1). It is characterised by retinal whitening, cotton-wool patches, and superficial retinal haemorrhages. Purtscher retinopathy has since been described in association with various types of trauma, including long-bone fracture, cephalic or thoracic compression, and crush injury (1). Cases such as acute pancreatitis, collagen vascular diseases, HELLP syndrome, renal failure, multiple myeloma and no relation to trauma have been reported with similar clinical manifestation (1). These cases are grouped together as Purtscher-like retinopathy.

Systemic lupus erythematous (SLE) is a disease that impacts multiple organ systems. It is a multifactorial disease of unknown aetiology in which significant immunological abnormalities have been identified. SLE can involve systemically and ocularly. About one third of patients with SLE have ocular manifestations (1). The eyelid, ocular adnexa, sclera, cornea, uvea, retina, and optic nerve can be affected by SLE. The most prevalent symptom is keratoconjunctivitis sicca. However, involvement of the retina, choroid, or optic nerve can result in visual loss. Ocular symptoms are linked to systemic disease activity and might appear as the first sign of SLE.

Here, we report a 10-year-old Chinese girl who presented with pyrexia of unknown origin in which finally diagnosed as SLE.

2 CASE REPORT

A 10-year-old Chinese girl was presented with lethargy for four weeks and intermittent fever for two weeks. The fever was associated with multiple rashes over the face and body. She was treated as pyrexia of unknown origin by the primary health care. She denied any blurring of vision, eye pain or redness. On examination, generally she was febrile with temperature of 39°C and stable other vital signs. There were presence of facial rash and discoid rashes involving both hands with generalised erythematous skin over the hernial region. However, there was no hepatomegaly, splenomegaly, or lymphadenopathy.

She was referred for eye assessment to rule out any ocular association of connective tissue disease. Ocular examination showed the visual acuity was 6/15 in the right eye and 6/30 in the left eye. Anterior segment examination was normal in both eyes. Fundus examination showed presence of generalised Purtscher-like retinopathy feature with multiple area of polygonal retinal whitening between the retinal arterioles and venules at posterior poles in both eyes (Figure 1). The retinal vessels were torturous bilaterally. Optic disc was pink with normal cup-disc ratio in both eyes. Optical coherence tomography (OCT) showed hyperreflectivity of the inner retina, as well as presence of cystoid macular oedema and minimal subretinal fluid in both eyes (Figure 2). As it requires good patient cooperation, fundus fluorescein angiography was not performed in this patient.

Blood investigation revealed leukopenia, anaemia, low complement (C) 3 (C3) and complement C4 with positive both antinuclear antibody (ANA) and anti-double-stranded DNA (anti-dsDNA) antibody. She was finally diagnosed as childhood-onset SLE by paediatric team. She was treated with oral mycophenolate mofetil (MMF) 1 g twice per day, oral prednisolone 5 mg daily, tablet hydroxychloroquine 200 mg daily and oral aspirin 75 mg daily. After 12 months of systemic therapy, the visual acuity improved to 6/9 in right eye and 6/7.5 in left eye. Fundus examination showed resolution of retinal whitening bilaterally. There was resolution of cystoid macular oedema and subretinal fluid on repeated OCT assessment. Systemically, there was also resolution of facial rash and discoid rashes. Patient was given a yearly follow-up at the eye clinic and regular review at paediatrics clinic.

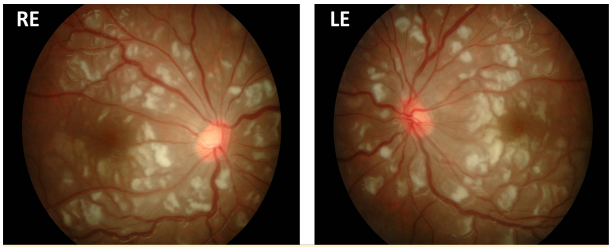

Figure 1. Fundus shows multiple areas of polygonal retinal whitening between the retinal arterioles and venules, with tortuous retinal vessels at the posterior pole in the right eye (RE) and left eye (LE)

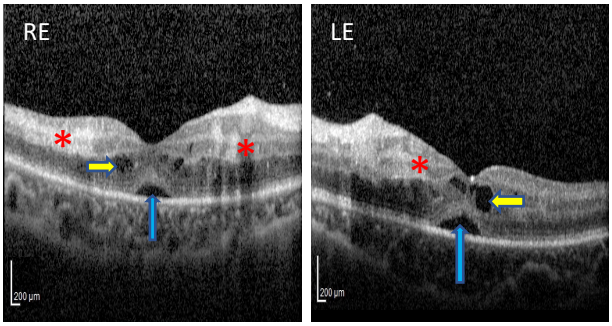

Figure 2. Optical coherence tomography (OCT) showed hyperreflectivity of the inner retina (*), presence of cystoid macular oedema (yellow arrow) and minimal subretinal fluid (blue arrow) in the right eye (RE) and left eye (LE)

3 DISCUSSION

Childhood-onset SLE is a rare condition, with a prevalence of 3.3-8.8 per 100.000 children and an incidence of 0.3-0.9 per 100.000 children-years (2). Childhood-onset SLE usually begins between the ages of 11 and 12 years old. The disease is uncommon in children under the age of five years old. Around 80% of patients with childhood-onset SLE are female, similar to adult-onset SLE (2).

SLE shares characteristics with many other (autoimmune) diseases as it is called the great mimicker. Fever, fatigue, anorexia, weight loss, alopecia and arthralgias are common nonspecific constitutional symptoms reported in SLE (2). The American College of Rheumatology (ACR) developed the SLE diagnostic criteria. It was based on 4 of 11 criteria, either at the present time or at some time in the past; malar rash, discoid rash, photosensitivity, oral ulcers, non-erosive arthritis, serositis, renal disorder, neurological disorder (seizures or psychosis), haematological disorder (anaemia, leucopenia, thrombocytopenia), immunological disorder (anti-DNA antibody, Anti-Smith [Sm] antibody and false positive Venereal Disease Research Laboratory [VDRL] testing) and presence of antinuclear antibodies (ANA). In this patient, she was diagnosed to have SLE based on the presence of facial rash, discoid rashes, haematological disorder (leukopenia and anaemia), immunological disorder (presence of anti-dsDNA) and ANA.

Retinal manifestation affects about 10% of SLE patients and can range from mild asymptomatic lupus retinopathy to severe blinding disease (3, 4). The most frequent retinal findings include cotton wool spots, retinal haemorrhages, and vascular

tortuosity (4, 5). Other reported posterior segment changes include retinal hard exudates, retinal vasculitis, retinal artery and/or vein occlusion, arteriolar narrowing, arteriovenous crossing changes, macular pigmentary mottling, retinal scarring, and macular infarction (6).

Severe vaso-occlusive retinopathy is a rare but well-known condition characterised by widespread retinal capillary non-perfusion, multiple branch retinal artery occlusions, ocular neovascularization, vitreous haemorrhage, tractional retinal detachment, neovascular glaucoma, and significant visual loss. Occlusions of the central retinal veins or arteries can happen alone or in pairs, and they can be unilateral or bilateral (6-8). In this patient, there was presence of Purtscher-like retinopathy, clinically manifest as polygonal retinal whitening between the retinal arterioles and venules in the both eyes. The retinal whitening is due to infarct of the capillary bed as a result of occlusion of small arterioles. In contrast, cotton wool spot is infarct of nerve tissue layer due to occlusion of the capillary (9).

The early detection and treatment of disease leads to a reduction in visual morbidity and mortality. Prompt systemic corticosteroids, steroid-sparing immunosuppressive medications, and biological agents are all part of the standard treatment. Despite proper treatment, visual acuity often did not restore back to normal (1). Early detection of the disease improved the prognosis and, as a result, the visual outcome can be restored back to normal. Continues monitoring of the disease activity is important in childhood-onset SLE since SLE in children required long term systemic medication that can lead to multiple complication related to the disease itself and also due to the side effect of the systemic medication.

4 CONCLUSION

Our case illustrates a rare but clinically significant ocular involvement in SLE in the form of Purtscher-like retinopathy. Hence, childhood-onset SLE presented with or without visual complaints must be evaluated thoroughly for Purtscher-like retinopathy. Early recognition and treatment lead to a better visual outcome.

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