Purtscher-like Retinopathy as a First Manifestation of Systemic Lupus Erythematosus

Authors.

Abstract

Purpose: To report a case of bilateral Purtscher-like retinopathy in a female patient as a first presenting sign which led to the diagnosis of systemic lupus erythematosus (SLE).

Case presentation: An otherwise healthy 21-year old female presented with bilateral rapid decrease in vision for one month. Fundus examination revealed multiple cotton wool spots and flame-shaped hemorrhages in both eyes. Optical coherence tomography displayed severe macular edema. Fundus fluorescein angiography showed multiple retinal arteriolar occlusions. Rheumatology was consulted for investigation. Work up for the patient confirmed the diagnosis of SLE. She was diagnosed with SLE and subsequently treated with corticosteroids and immunosuppressive medication.

Conclusion: Although vaso-occlusive retinopathy is a rare presentation of SLE, it is important to suspect the diagnosis of SLE in a patient with such presentation, in order to promptly initiate proper treatment.

Background

Systemic lupus erythematosus (SLE) is a complex autoimmune inflammatory disease of unknown etiology. It is characterized by involving multiple organ systems, often with a relapsing and remitting clinical course. Almost one third of SLE patients manifest an array of ocular manifestations. The most common manifestation is SLE retinopathy. The retinopathy typically presents with cotton wool spots with or without flame-shaped hemorrhages [(1)]. SLE retinopathy is usually manifested in patients with active disease, and is more common when associated with antiphospholipid syndrome. The presence of antiphospholipid antibodies, in isolation or in association with SLE, increases the risk of retinopathy [(2)]. PLR has been reported to be associated with SLE. This report describes the development of PLR in a young female as a first manifestation of SLE

Case Presentation

A 21-year-old female, presented to King Abdulaziz University Hospital (KAUH) emergency department with rapid painless decreased vision in both eyes for one month. Ophthalmic examination showed a best-corrected visual of counting fingers at 3 feet for both eyes. The pupils were found to be regular and reactive with no afferent pupillary defect in both eyes. External eye examination was normal. Slitlamp examination revealed an unremarkable anterior segment. Intraocular pressure was within normal range. Fundoscopy showed a clear vitreous and the presence of large, multiple cotton wool spots and flame-shaped hemorrhages in both eyes. The optic disk was normal (figure 1). Fundus fluorescein angiography revealed multiple arteriolar occlusions and large areas of capillary drop out (figure 1). Optical Coherence Tomography (OCT) displayed severe macular edema of 826 μm and 853 μm for the right and left eye, respectively (figure 2). The diagnosis of bilateral Purtscher-like retinopathy (PLR) was made and an underlying systemic vasculitis entity was suspected. The patient was referred for rheumatology consult.

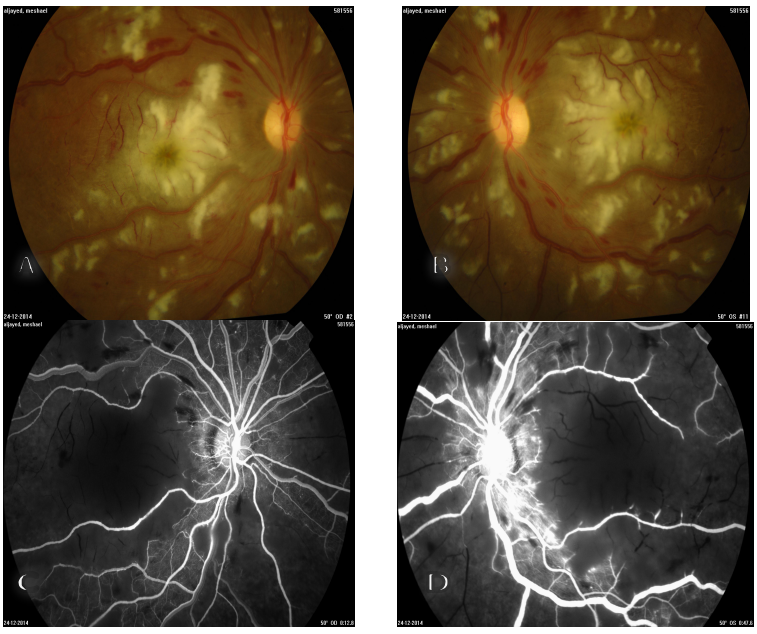


Figure 1. 21-year old woman had decreased vision in both eyes for one month. (A, B) Fundus photographs show multiple cotton wool spots and flame shaped hemorrhages. (C, D) Fluorescein angiography shows multiple arteriolar occlusions and large areas of capillary drop-out.cc

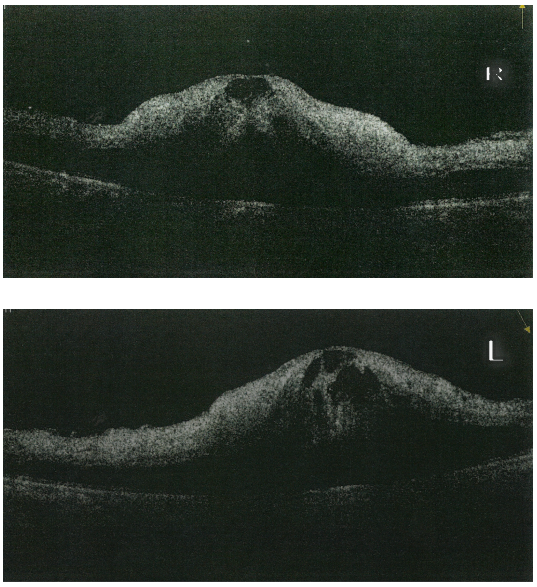


Figure 2. Optical Coherence Tomography of a woman with systemic lupus erythematous. OCT displayed severe macular edema of 826 µm and 853 µm for the right and left eye respectively.

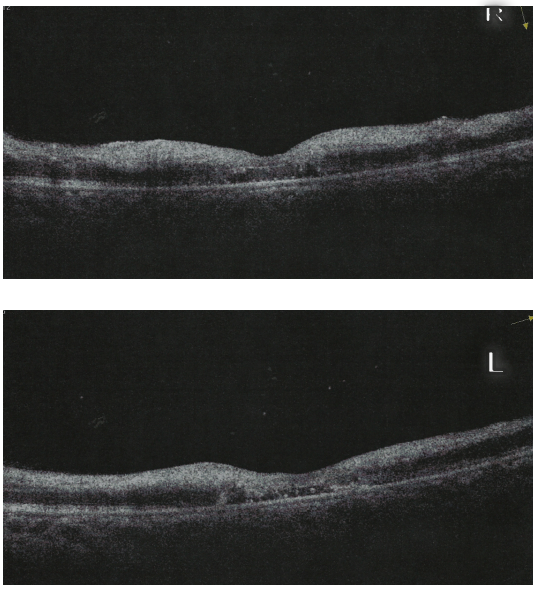


Figure 4. Optical Coherence Tomography of a woman with PRL associated with systemic lupus erythematous after treatment. OCT showing decreased retinal thickness and macular edema to 226 µm and 169 µm of both right and left eye, respectively.

A detailed medical history revealed that the patient had contracted persistent low-grade fever, rigors, night sweats, anorexia, fatigue, and complaints of oral ulceration and erythema over the cheeks, suggesting malar rash, two months prior to presentation. She had an unremarkable past medical history and was not on any medication. On examination she was 60 inches tall and weighted 110 lb. She was found to have generalized lymphadenopathy and subungual pre-gangrene later affirmed as a sign of vasculitis. The remainder of the physical examination was unremarkable with no signs of skin manifestations, mucocutaneous ulcers, or arthritis.

Initial laboratory investigations showed evidence of pancytopenia; leukocytes 1.9x10 ^9/L, neutrophils 1.4x10 ^9/L hemoglobin 81 g/L, reticulocytes 1.1%, and platelets 71x10^9/L. Mid-stream urine and dipstick were significant for +2 protein, +2 hemoglobin, 42 WBCs, and 51 RBCs with no growth recorded. 24-hour urine sample revealed total protein loss of 0.43 g/L, serum albumin 23 g/L. CT demonstrated few tiny mediastinal lymph nodes. Axillary lymph node biopsy reported lymphoid hyperplasia, no malignant cells.

Hematologic workup revealed antinuclear antibody titer levels of >1:2560 (N<1180) with speckled pattern, anti-DNA antibodies at 1734 i.u/ml (N<200 i.u/ml), and elevated levels of anticardiolipin antibodies (ACA) IgM 31 (N< 18 MPL), ACA IgG 23 (N< 14 GPL). C3 and C4 were decreased to 0.178 and 0.041 respectively. Ultrasound guided kidney biopsy was obtained and confirmed class IV lupus nephritis with minimal disease activity, plus very early membranous changes class V.

Based on the American College of Rheumatology 1982 revised criteria for diagnosing SLE, a diagnosis Systemic Lupus Erythematosus with moderately elevated antiphospholipid antibody was made. She was treated with 20 mg intravenous methylprednisilone to b tapered off for 3 months according to her response. She was discharged on 50 mg prednisone for 6 days, to be tapered down by 5 mg every week and maintained at 10 mg once daily for 2 months, 500mg of mycophenolate mofetil, twice, daily for 5 days, 200 mg hydroxychlorouquine sulphate, and warfarin 2 mg for 3 months to be increased to 1 mg, within 1 month and to continue this does for at least 2 years.

After one-month, her follow up at the ophthalmology clinic at KAUH recorded an improved visual acuity to 20/400 and 20/200 for the right and left eyes respectively. Fundus examination revealed decrease in number and size of cotton wool spots and a pale optic nerve head (Figure 3). OCT was performed showing decreased retinal thickness and resolved macular edema to 226 µm, 169 µm of both right and left eye respectively. (Figure 4) Additional follow up with the eye clinic was recommended after 8 to 10 weeks of treatment for monitoring. No additional medication was prescribed.

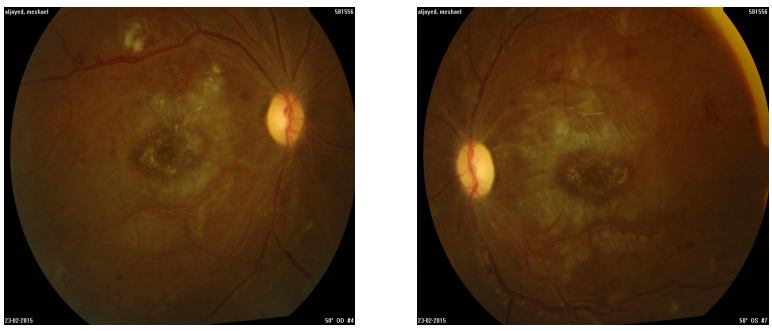


Figure 3. Fundus Photograph of a woman with systemic lupus Erythematosus retinopathy one-month post treatment. Fundus photograph shows decrease in number and size of cotton wool spots and resolved macular edema bilaterally.

Discussion

Depending on 1997 update of the 1982 American College of Rheumatology Revised Criteria for Classification of Systemic Lupus Erythematosus [3], retinopathy is not included in the diagnostic parameters; it is a well-known complication of SLE with an incidence of 29% in patients with active disease [1].

Purtscher retinopathy (PR) generally occurs as a result of trauma or thoracic compression. A funduscopic picture virtually identical to that of PR may occur with non-traumatic causes and labeled Purtscher-like retinopathy. The most frequently associated is acute pancreatitis. Autoimmune disorders such as SLE, dermatomyositis, and scleroderma have also been reported. [4]

The characteristic ophthalmic findings in PLR include multiple patches of superficial retinal whitening and retinal hemorrhages surrounding the optic nerve head, which usually appears normal. Fluorescein angiography usually demonstrates leakage of dye from retinal arterioles, capillaries and venules, in patients with relatively mild PLR. In this case, evidence of arteriolar obstruction and dye leakage was noted, which is characteristic of a severe form of PLR [5].

This case is distinguished for the manifestation of Purtscher-like retinopathy as a first manifestation of SLE. Chang-Wu et al reported 8 cases of Purtscher-like retinopathy in patients previously diagnosed with SLE in an observational case series [6]. The ophthalmic complication was contributed to central nervous lupus and highly active disease (>20). Our patient displayed disease activity of 41 according to the SLE Disease Activity Index (SLEDAI). To our knowledge, few others reported PLR associated with SLE [6-10], all of whom recorded good visual-acuity prognosis following corticosteroid treatment. However, patients with present antiphospholipid antibodies had a more severe form of retinopathy and carried a worse visual prognosis.

The pathogenesis of PLR is still uncertain. It has been proposed that complement activation and leukoembolization by leukocytary aggregation with secondary lymph extravasation, is implicated in the occurrence of PLR. [7, 11, 12].

In conclusion, we herein report a young female patient who presented with severe Purtscher-like retinopathy, which led to the diagnosis of SLE. This case highlights the fact that ocular manifestation could precede occurrence of a serious manifestation of SLE and the role ophthalmologists could play in the diagnosis of SLE.

Acknowledgements:

We are grateful to the deanship of the scientific research of king Saud University for their support in writing this scientific article.

Conflict of interest:

The authors whose names are listed immediately below certify that they have NO affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

References:

1. Palejwala NV, Walia HS, Yeh S. Ocular Manifestations of Systemic Lupus Erythematosus: A Review of the Literature. Hindawi Publishing Corporation Autoimmune Diseases 2012; 2012(290898):9.
2. Au U, O'Day J. Review of Severe Vaso-occlusive retinopathy in SLE and the Anti-Phospholipid Syndrome; Associations, Visual Outcomes, Complications and Treatment. Clin Experimint Ophthalmol 2004;32;87-100.
3. Hochberg MC: Updating the american college of rheumatology revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum 1997, 40:1725.
4. Miguel AI, Henriques F, Azevedo LF, Loureiro AJ, Maberley DA. Systematic Review of Purtsher's and Purtsher-like Retinopathies. Eye (Lond) 2013;27(1):1-13.
5. Gass JD. Sterioscopic Atlas of Macular Diseases: Diagnosis and Treatment. Volume 2. Third edition.
6. Wu C, Dai R, Dong F, Wang Q. Purtscher-like Retinopathy in Systemic Lupus Erythematosus. American J of Ophthalmol 2014; 158(6):1335-1341.
7. Sellami D, Ben Zina Z, Jelliti B, Abid D, Feki J, Chaabouni M. Purtscher-like Retinopathy in Systemic Lupus Erythematosus. Two Cases. J Fr Ophthalmol 2002; 25(52-55).
8. Kunavisarut P, Pathanapitoon K, Rothova A. Purtscher-like Retinopathy Associated with Systemic Lupus Erythematosus, Posted Online (doi: 10.3109/09273948.2014.932816) 2014.
9. Prabhu. Loss of Vision Following Fever in a Young Female. AECS Illumination;14-16.
10. Cruz FM, Chan I, Tee C, Gulay C, Dans L, Otadoy-Augustin J, Recto M. Retinopathy in a Patient with Systemic Lupus Erythematosus. Philipp J Ophthalmol. 2009;34(2):66-69
11. Pedro G, Vinay A. Purtscher Retinopathy and Purtscher-like Retinopathy. Eyewiki.aao.org. 2014.
12. Agrawal A, Mckibbin M. Purtscher's Retinopathies: A Review. Surv Ophthalmol 2006;51:129-136