Bilateral central retinal artery occlusion - A catastrophic presentation of systemic lupus erythematosus

Somya Ish, Deepa Sharma, Rahul Verma, Sudha Kumari, Himanshu Garkoti

Abstract:
Systemic lupus erythematosus (SLE) is a multisystem autoimmune disorder. Here, we present a rare case of a middle-aged male, diagnosed with SLE, manifesting as bilateral central retinal artery occlusion (CRAO). Severe ocular complications such as CRAO may occur during an acute flare or even early in the disease process. It is important to recognize this potentially devastating complication.

Keywords:
Central retinal artery occlusion, rare, systemic lupus erythematosus

Introduction
Systemic lupus erythematosus (SLE) is a multisystem autoimmune disorder affecting females much more frequently than males.[1] Ocular complications are found in up to one-third of patients with SLE that may affect any structure of the eye and adnexa.[2,3] We present a rare case of a middle-aged male, diagnosed with SLE, manifesting as bilateral central retinal artery occlusion (CRAO).

Case Report
A 52-year-old male patient came with a complaint of sudden painless loss of vision in the left eye for 1 week and the right eye for 1 day. He was a diagnosed case of SLE according to the Systemic Lupus International Collaborating Clinics classification[4] with mucocutaneous manifestations, including facial rash [Figure 1], oral ulcers, anemia (hemoglobin level of 10 g/dl), and photophobia for 2 months.

Antinuclear antibodies profile was positive for autoantibodies against SmDI, PO, SS-A/Ro60, SS-A Ro52, and SS-B/La antigens. The antiphospholipid antibody profile of the patient was negative. He was on tablet hydroxychloroquine 400 mg once a day and injection methylprednisolone 80 mg intramuscular once per week for the past 1 month.

On detailed ocular examination, he had no perception of light in both the eyes. The conjunctiva was normal, the cornea was clear, and anterior chamber depth was Van Herick’s Grade IV. The pupil of both the eyes was mid-dilated and not reacting to both direct and consensual light reflexes. Fundus examination of the right eye revealed pale disc and retina with a cherry-red spot at the fovea with arteriolar attenuation suggestive of CRAO [Figure 2]. On fundus examination of the left eye, the disc was edematous and pale, and the whole retina was pale with attenuation of both vein and arterioles suggestive of CRAO with old vasculitis [Figure 3].

The patient was given immediate ocular massage in both eyes and intravenous 350 ml of 20% mannitol. Intravenous...
Ish, et al.: CRAO in SLE

Discussion

Ocular involvement in SLE can range from all manifestations, including keratoconjunctivitis sicca, scleritis, uveitis, ischemic optic neuropathy, and occlusion of retinal vessels. The posterior segment findings, particularly the retinal signs in SLE, reflect the severity of systemic inflammation and may indicate the inadequate control of the systemic disease. CRAO is an ophthalmic emergency presenting as profound acute visual loss that may be irreversible. The severe form of vaso-occlusive retinopathy resulting from sudden occlusion of a bilateral central retinal artery is a rare manifestation. Immune complex-mediated “vasculitis” has often been described as the pathology for this severe vaso-occlusive retinopathy, but the precise nature of these occlusive lesions remains unclear. Histologically, fibrinoid change with thrombosis in the vessel walls has been postulated without evidence of inflammation because of the relative inaccessibility of the retinal vessels for histopathological study, this is also sparsely discussed in the literature. Small retinal vessels are more often involved than large vessels and arteries more than veins.

This is a potentially visually devastating form of retinopathy in SLE, resulting in a visual loss in about 80% of affected patients. Despite giving systemic as well as local treatment, the vision of the patient could not be recovered.

Conclusion

SLE is a multisystem disease which requires the collaboration between rheumatologists, ophthalmologists, and nephrologists, to provide early effective treatment and prevent complications. Severe ocular complications such as CRAO may occur during an acute flare or even early in the disease process. It is important to recognize this potentially devastating complication by maintaining a high index of suspicion for the same.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

methylprednisolone 1 g per day was given for 3 continuous days along with hyperbaric oxygen, but there was no improvement in vision.
References

1. Pons-Estel GJ, Alarcón GS, Scofield L, Reinlib L, Cooper GS. Understanding the epidemiology and progression of systemic lupus erythematosus. Semin Arthritis Rheum 2010;39:257-68.

2. Read RW. Clinical mini-review: Systemic lupus erythematosus and the eye. Ocul Immunol Inflamm 2004;12:87-99.

3. Davies JB, Rao PK. Ocular manifestations of systemic lupus erythematosus. Curr Opin Ophthalmol 2008;19:512-8.

4. Petri M, Orbai AM, Alarcón GS, Gordon C, Merrill JT, Fortin PR, et al. Derivation and validation of the systemic lupus International Collaborating Clinics Classification Criteria for Systemic Lupus Erythematosus. Arthritis Rheum 2012;64:2677-86.

5. Sivaraj RR, Durrani OM, Denniston AK, Murray PI, Gordon C. Ocular manifestations of systemic lupus erythematosus. Rheumatology (Oxford) 2007;46:1757-62.

6. Fouad el A, Hanane M, Mounir B, Rachid Z, Karim R, Abdelber O. Severe ischemic retinopathy in a patient with systemic lupus erythematosus without antiphospholipid syndrome: A case report. Saudi J Ophthalmol 2015;29:169-71.

7. Varma DD, Cugati S, Lee AW, Chen CS. A review of central retinal artery occlusion: Clinical presentation and management. Eye (Lond) 2013;27:688-97.

8. Hua L, Patel K, Corbett JJ. Bilateral central retinal artery occlusion in a patient with systemic lupus erythematosus. J Stroke Cerebrovasc Dis 2015;24:e139-41.

9. Gold D, Feiner L, Henkind P. Retinal arterial occlusive disease in systemic lupus erythematosus. Arch Ophthalmol 1977;95:1580-5.

10. Ermakova NA, Alekberova ZS, Kosheleva NM, Reshetniak TN. Characteristics of retinal vascular involvement in systemic lupus erythematosus. Vestn Oftalmol 2001;117:21-4.

11. Au A, O’Day J. Review of severe vaso-occlusive retinopathy in systemic lupus erythematosus and the antiphospholipid syndrome: Associations, visual outcomes, complications and treatment. Clin Exp Ophthalmol 2004;32:87-100.