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CLINICAL APPROACH TO VASO-OCCLUSIVE RETINOPATHY SYSTEMIC LUPUS ERYTHEMATOSUS: A CASE SERIES

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ABSTRACT

Background: Two patients reported experiencing episodes of blurred vision in their left eye, occurring over the last one months, accompanied by headaches. Both individuals had a medical history of Systemic Lupus Erythematosus (SLE).

Case Presentation: Two cases of blurred vision on the patients with diagnosis of SLE. From the ophthalmology examination on two cases was found hemorrhage, exudate, cotton wool spots and macular edema. They were diagnosed vaso-occlusive retinopathies SLE. Diagnosis was confirmed in accordance with the European League Against Rheumatism (EULAR) and the American College of Rheumatology (ACR) criteria, which necessitate the presence of specific clinical symptoms and laboratory findings. Diagnosis requires a minimum antinuclear antibody (ANA) titer of 1:80 on HEp-2 cells or an equivalent positive test at least once. The 22 'additive weighted' classification criteria were applied, involving immunologic domains (antiphospholipid antibodies, complement proteins, and SLE-specific antibodies) alongside seven clinical domains (constitutional, hematological, neuropsychiatric, mucocutaneous, serosal, musculoskeletal, and renal). Each criterion is assigned a point value ranging from two to ten. A diagnosis of SLE is made if at least one clinical criterion is met and the total reaches ten or more points.

Conclusion:.This case series highlights atypical "lupus retinopathy," where severe vaso-occlusive events may cause blindness and require urgent treatment—including immunosuppression, anticoagulation, and addressing hypercoagulability. Clinicians should suspect undiagnosed antiphospholipid syndrome in SLE patients, as it can lead to major vascular occlusions. The goal is to raise clinician awareness, especially among ophthalmologists, of these ocular manifestations and their management.

Keywords: *Vasso-Occlusive, Retinopathy, Systemic Lupus Erythematosus*

BACKGROUND

Systemic lupus erythematosus (SLE) is an autoimmune disorder of unknown origin, marked by the presence of circulating autoantibodies targeting various cell nuclear components, leading to immune-mediated tissue damage. Clinical manifestations are diverse, with musculoskeletal and skin symptoms being the most prevalent. Retinopathy associated with SLE has been well documented, with a recognized correlation between antiphospholipid antibodies, retinopathy, and central nervous system involvement. While most cases have a favorable visual prognosis, a small subset of patients may develop severe vaso-occlusive retinopathy, often resulting in poor visual outcomes. The pathogenesis of this severe form has been linked to immune complex-mediated vasculitis; however, recent findings suggest that thrombosis associated with antiphospholipid syndrome plays a significant role. This raises treatment considerations regarding anticoagulation versus immunosuppression. A literature review of previously reported cases was conducted to assess current knowledge on pathogenesis, treatment, visual outcomes, and associated factors.[1-8]

CASE PRESENTATION

Case 1:

A 31-year-old female patient complained of blurred vision in her left eye since 1 month ago. The patient's vision was gradually getting blurrier. Complaints such as red eyes, watery eyes, headaches, nausea and vomiting, double vision, closed vision on one side, seeing like seeing a rainbow, flying objects or closed curtains were believed by the patient. The patient had a history of SLE since 2 months ago and started immunosuppressive therapy. From the ophthalmology examination, VOD 6/21 PH (6/7.5) and VOS <1/60 were obtained. The anterior segment examination was found to be within normal limits. The posterior segment examination found round papillae, firm boundaries, normal red color, C/D ratio 0.3, a:v 2:3, hard exudate on the macula, flame-shaped hemorrhage in 1 quadrant, hard exudate in 2 quadrants and cotton wool spots in 4 quadrants in the left eye. While the posterior segment examination of the right eye was found to be within normal limits. On OCT examination, a dome-shaped hyporeflectivity image was obtained above the RPE layer with foveal depression that disappeared in the left eye. While in the right eye within normal limits. The decreased vision in the eye is in accordance with the findings of macular edema in the right eye as seen from the OCT examination. In this patient, the results of laboratory tests supported, namely, Anti DsDNA: reactive, ANA: 1/160 results and platelets 156,000 mg/dL. The patient was treated by a rheumatologist with Cyclosporine 100 mg per day and methyl prednisolone oral 32 mg per day. In this case, based on the follow-up results after one month, progress was found in the form of improved vision to 6/60 and the condition of the macula improved.

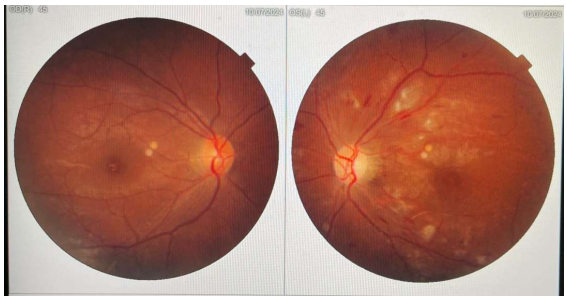

Figure 1: Funduscopy Examination (Case 1)

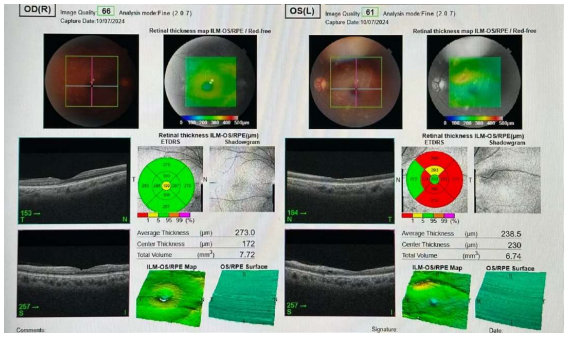

Figure 2: Macula OCT Examination (Case 1)

Case 2:

An 18-year-old male patient complained of blurred vision in his left eye since 1 month ago. The patient's vision was felt to be getting blurrier slowly. Complaints such as red eyes, watery eyes, headaches, nausea and vomiting, double vision, closed vision on one side, seeing like seeing a rainbow, flying objects or being covered by curtains were denied by the patient. The patient had a history of SLE since 2 months ago and started immunosuppressive therapy. The patient was then consulted to the neuroophthalmology subdivision. On initial examination, an ophthalmological examination found VOD 6/9 PH 6/6 and VOS <1/60. On examination of the anterior segment, it was found to be within normal limits. On examination of the posterior segment of the left eye, round papillae were found, with clear borders, normal red color, C/D ratio was difficult to assess, a:v 2:3, peripapillary splinter hemorrhage, arteriovenous dilation in 4 quadrants, intra-retinal hemorrhage in 2 quadrants, hard

exudates in 2 quadrants, cotton wool spots in 4 quadrants, perivascular sheathing, while on examination of the posterior segment of the right eye, perivascular sheathing was also found. On OCT examination, a dome-shaped hyporeflectivity image was obtained above the RPE layer with foveal depression that disappeared in the left eye. While in the right eye it was within normal limits. The decreased vision in the eye was in accordance with the findings of macular edema in the right eye as seen from the OCT examination. In this patient, laboratory examination results were obtained that supported, namely, Anti DsDNA: reactive, ANA: 1/3200 and platelets 352,000 mg/dL. She was also referred to the neurology department, where the diagnosis of suspected Neuropsychiatric SLE (NPSLE) was made. The patient was treated by a rheumatologist with Cyclosporine 100 mg per day, methyl prednisolone (IVMP) 125mg per day and anti platet 80mg per day. In this case, based on the follow-up results after 2 weeks, progress was found in the form of improved vision to 3/60 and the condition of the macula improved.

Figure 3: Funduscopy Examination (Case 2)

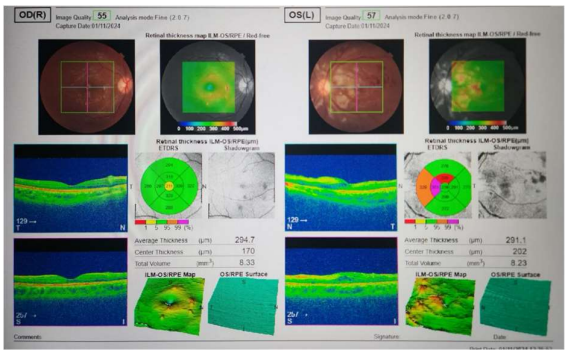

Figure 4: Macula OCT Examination (Case 2)

DISCUSSION

The literature describes a milder form of SLE retinopathy, with reported incidences ranging from 3.3% to 28.1%, increasing with the severity of systemic disease. Common findings include cotton wool spots, retinal hemorrhages, and optic disc edema, which correlate with SLE disease activity. Despite being a marker for poor prognosis regarding survival, this variant typically preserves visual acuity. The condition is considered an immune complex-mediated vasculopathy, although reports of direct immune complex deposition in retinal vessels are limited. Instead of being a true inflammatory vasculitis, retinal vascular disease is believed to result from fibrinoid degeneration and necrosis of vessel walls, often without ocular inflammation. Factors such as accelerated atherosclerosis due to hypertension, corticosteroid use, and dyslipidemia may contribute to this benign retinopathy. Fluorescein angiography in patients with SLE, even in the absence of visual symptoms, has shown evidence of retinal capillary dilatation and microaneurysms, indicating an underlying vascular abnormality that is frequently subclinical. In contrast, severe occlusive retinopathy is rare and typically affects smaller retinal arteries. This form often results in visual impairment, with vascular occlusion occurring in vessels of varying sizes and histologically presenting as non-inflammatory bland thrombosis. Approximately 30% of SLE patients have antiphospholipid antibodies, and the

antiphospholipid syndrome is observed in 50-70% of those with both SLE and these antibodies after two decades of follow-up. Research indicates a relationship between SLE disease activity and antiphospholipid antibodies, suggesting that treatment of the underlying systemic disease can diminish both the presence and levels of these antibodies. Key risk factors for thrombosis include prior thrombotic events and high titers of antiphospholipid antibodies. However, once a thrombotic event occurs, the risk of subsequent events remains elevated indefinitely, regardless of the presence of these antibodies.[9-20]

CONCLUSION

Severe vaso-occlusive retinopathy, though rare, causes significant visual loss in up to 80% of cases, with neovascularization or vitreous hemorrhage in about 40%. Its pathogenesis may involve antiphospholipid antibodies and microthrombosis, linking it to antiphospholipid syndrome. As this condition is often irreversible, early treatment to prevent neovascular complications is crucial. Anticoagulation may help prevent further thrombosis in patients with antiphospholipid antibodies. This series underscores the need for prompt recognition and management of atypical lupus retinopathy through immunosuppression, anticoagulation, and assessment of systemic hypercoagulability. Ophthalmologists should remain alert for undiagnosed antiphospholipid syndrome in SLE patients due to its potential for causing severe vascular occlusion

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