
CASE REPORT

NON-NECROTIZING DIFFUSE ANTERIOR SCLERITIS AS AN ISOLATED OCULAR MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS: A CASE REPORT**Kara Citra Kalandra¹, Rifna Lutfiamida²**¹ Dr. Suyoto Hospital, Ministry of Defense Republic of Indonesia²KMN EyeCare, Jakarta

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ABSTRACT

Introduction: Systemic lupus erythematosus (SLE) is an autoimmune disorder with heterogeneous clinical manifestation in numerous organ systems, including the eye. Ocular involvements have been reported in up to one-third of patients with SLE, although scleritis is considered a less common manifestation of the disease found in only about 1% of cases

Case Report: A 46-year-old female patient presented with a painful right eye for 1 week. She reported having the same symptom last year which resolved without treatment. There was no known history of systemic immune-mediated diseases and no prior ocular trauma. Ophthalmological examination revealed dilatation of deep scleral vessels on the right eye. The patient tested positive for the antinuclear antibody and anti-double-stranded deoxyribonucleic acid tests. She was diagnosed with non-necrotizing diffuse anterior scleritis caused by SLE and was treated with topical prednisolone acetate and systemic methylprednisolone. After 1 week of treatment, the patient showed an improvement in ocular signs and symptoms.

Discussion: Scleritis is a rare inflammatory disease usually associated with systemic immune-mediated diseases such as SLE. It can cause significant visual loss and may be life-threatening. We reported a non-necrotizing diffuse anterior subtype of scleritis presented as an isolated manifestation in SLE, which benefited from early diagnosis and treatment of corticosteroids.

Conclusion: Scleritis may present as an early or isolated manifestation of systemic immune-mediated disease. Therefore, a comprehensive examination is of the essence to exclude multisystem disease and treat the underlying cause even when there is no systemic manifestation.

Keywords: Scleritis, Diffuse Anterior Scleritis, Systemic Lupus Erythematosus

INTRODUCTION

Scleritis is a painful and vision-threatening inflammation involving the sclera and its adjacent structures. Scleritis can be isolated in the eye, but more commonly as a manifestation of systemic disease in up to half of affected individuals.^{1,2} Scleritis is often associated with systemic immune-mediated diseases and can also be an early manifestation of a systemic connective tissue or vasculitic diseases, such as rheumatoid arthritis, Wegener's granulomatosis, or systemic lupus erythematosus.^{3,4}

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disorder with heterogeneous clinical manifestation in numerous organ systems, including the eye. The prevalence of SLE varies across global regions even within subpopulations, ranging from 13 to 7,713.5 per 100,000 individuals.⁵ It typically *affects females* at far greater rates *than males with a ratio of 9 to 1*, predominantly the productive age population.⁶ It is also one of the leading causes of death in young women.⁷ Ocular involvements have been reported in up to one-third of patients with SLE and may affect various parts of the visual pathway, the most common being keratoconjunctivitis sicca (KCS).⁸⁻¹⁰ Scleritis is considered a less common manifestation of the disease, found in only about 1% of cases.⁸

Diagnosis of scleritis requires a high index of clinical suspicion, especially outside the context of an active systemic manifestation. It is important to exclude multisystem diseases and treat the underlying cause, not only as it helps to control the ocular inflammation, but also to halt the relentless progression of the systemic destructive processes. This case report presents an uncommon case of non-necrotizing diffuse anterior scleritis caused by SLE without any signs of other organ involvement.

CASE ILLUSTRATION

A 46-year-old female patient presented with recurrent painful red eye in the right eye (RE) for 1 week. She described the pain as periorbital fullness, gradually worsened as a deep severe radiating ache. It increases with eye movements and responds poorly to painkillers, causing difficulty in sleeping at night. She admitted to having been under a lot of stress and eating an unhealthy diet. Her vital signs and physical examination were normal. The patient denied having fatigue, joint pain, fever, rashes on the cheeks, or any other complaints besides the ocular symptoms. There was no known history of past ocular or systemic diseases and no prior ocular trauma.

Initial ophthalmological examination revealed VA on both eyes was 6/6. Examination of the RE with daylight and slit lamp revealed scleral edema and congestion without signs of necrosis. Mild photophobia was elicited during pupil and extraocular motility testing. Intraocular pressure and funduscopy examination were normal in both eyes. The results of complete blood count, chest x-ray, and brain computed tomography (CT) were within normal limits. This patient tested positive for Antinuclear Antibody Test (ANA Test) and Anti-double stranded DNA (Anti-dsDNA) indicative of SLE. Her Tuberculin Skin Test (TST) and Interferon-Gamma Release Assays (IGRA) tests were negative.

She was diagnosed with non-necrotizing diffuse anterior scleritis caused by systemic lupus erythematosus (SLE) and was treated alongside an internist with topical methylprednisolone acetate 4 administrations a day, and systemic methylprednisolone with a starting dose of 1 mg/kg/day. Aside from medical treatment, the patient was also asked to avoid stress and improve her dietary intake. After 1 week of treatment, the patient showed an improvement in ocular signs and symptoms. There was no recurrence and both eyes were no longer painful. Dilatation of scleral vessels has significantly decreased and her vision remains unimpaired.

Table 1. Ophthalmological examination at the initial visit and after one week of treatment

	Initial Visit	One week of treatment
RE	<ul style="list-style-type: none"> ▪ VA: 6/6 ▪ Inflammation: 3+ Severe scleral inflammation with diffuse significant redness of sclera and edema with no signs of necrosis ▪ Pain: Severe 	<ul style="list-style-type: none"> ▪ VA: 6/6 ▪ Inflammation: 1+ Mild scleral inflammation with diffuse mild dilation of deep episcleral vessels ▪ Pain: None
LE	<ul style="list-style-type: none"> ▪ VA: 6/6 ▪ Inflammation: - ▪ Pain: None 	<ul style="list-style-type: none"> ▪ VA: 6/6 ▪ Inflammation: - ▪ Pain: None

Note. Inflammation grading source in accordance to Sen HN, Sangave AA, Goldstein DA, Suhler EB, Cunningham D, Vitale S, Nussenblatt RB. A standardized grading system for scleritis. Ophthalmology. 2011 Apr;118(4):768-71

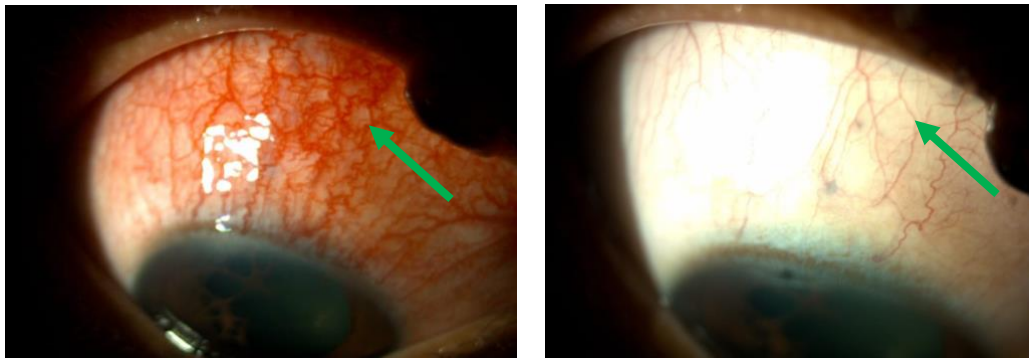


Figure 1. Slit lamp examination of initial and after one week of treatment showed improvement in the dilatation of scleral vessels

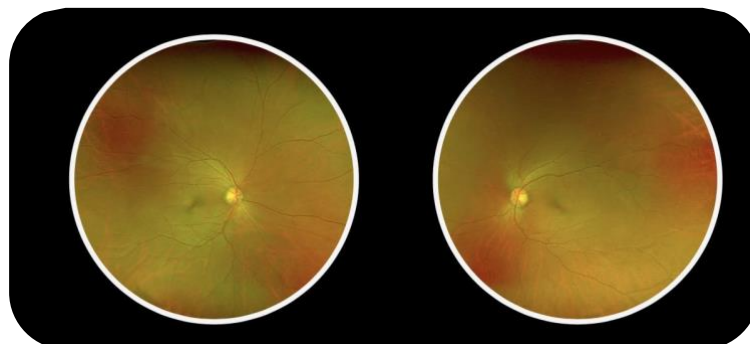


Figure 2. Initial funduscopy examination showed no vasculitis or optic disc edema in both eyes

DISCUSSION

Scleritis is a rare inflammatory disease involving the sclera and its adjacent structures. It may cause severe radiating pain and prompt action is needed to prevent significant visual loss. The earliest classification proposed by Watson and Hayreh is still being used until this day, differentiating scleritis into anterior and posterior forms. Anterior and posterior scleritis is distinguished by the involvement of the sclera beyond the equator to the insertion of the rectus muscles. Anterior scleritis is more commonly found and can further be divided into diffuse, nodular, and necrotizing forms.^{11,12}

Scleritis can be idiopathic or caused by either immune-mediated diseases or infectious pathogens, although up to half of the cases have an identifiable systemic disease that is presumed to be causally related.² It can also be the early manifestation of a systemic connective tissue or vasculitic diseases, some of which are life-threatening.^{3,4,13} The detection of such diseases may also be a sign of poor general and ocular prognosis because it indicates serious systemic and ocular complications.³ Therefore, patients with scleritis require a systemic evaluation to exclude multisystem diseases and treat the underlying cause. This should start with a thorough medical history, an extensive physical and laboratory examination in addition to a full ophthalmology examination.

Systemic lupus erythematosus is an autoimmune disorder with heterogeneous clinical manifestation in numerous organ systems, and one of the vasculitic diseases that may cause scleritis. It has previously been reported as a less common cause of scleritis than rheumatoid arthritis or Wegener's granulomatosis, found in only about 1% of cases.⁸ However, it is worth noticing as it is one of the leading causes of death in productive-aged women.⁷ The pathogenesis of SLE is largely unknown, previous studies have suggested that the manifestation of this disease is the result of several environmental, hormonal, and nutritional factors that, in subjects with a genetic predisposition contributed to impaired immune responses activation and subsequent inflammation.¹⁴

In this case, the patient presented with a chief complaint of severe pain of the RE that increases with eye movements and responds poorly to painkillers, symptoms commonly found in scleritis. However, the patient had not experienced fatigue, joint pain, fever, or any other symptoms often associated with SLE. There was also no known history of past ocular or systemic diseases and no prior ocular trauma. Ocular involvements are usually adjacent to the typical clinical features associated with SLE and an isolated unilateral ocular manifestation is considered uncommon, although some cases have been reported both in adults and children.^{15,16}

Scleritis in patients with SLE usually presents as anterior diffuse or nodular, whilst anterior necrotizing and posterior scleritis are considered uncommon.⁸ This is in accordance with our finding of anterior diffuse scleritis without signs of necrosis, and normal fundoscopic examination. The patient reported being under a lot of stress recently, which we presumed to be the trigger that set off the illness since she does not take any regular medication and is rarely exposed to ultraviolet rays. In addition, stress has previously been reported as a trigger of disease onset and flares in up to 60% of SLE cases.^{17,18}

After reviewing medical history, clinical manifestation, and diagnostic procedure findings, we diagnosed this patient with unilateral non-necrotizing diffuse anterior scleritis as an isolated manifestation of SLE. Diagnosis in this patient is based on ocular findings supplemented by a positive ANA and Anti-ds-DNA tests indicative of SLE, after excluding alternative diagnoses. Although according to the American College of Rheumatology (ACR) and *Systemic Lupus International Collaborating Clinics (SLICC)* classification criteria of SLE, our patient falls within ‘‘probable’’ SLE, as she does not have any typical clinical features and no other sign of inflammation outside the eyes.¹⁹

This patient was then treated alongside an internist with topical methylprednisolone acetate 4 administrations a day, and systemic methylprednisolone with 40 mg as an initial dosage. Aside from medical treatment, the patient was also asked to avoid stress and improve her dietary intake. Although it may not cause or trigger SLE flares, having a well-balanced diet with restrictions on carbohydrates and protein in addition to taking nutritional supplements (*i.e.*, vitamins, minerals, and polyphenols) is beneficial for SLE patients to help minimize inflammation, prevent adverse effects of medication, and improve the patient's well-being.²⁰ After 1 week of treatment, the patient showed an improvement both in symptoms and ocular examinations. She has been compliant with the therapy and there have not been any recurrences nor any systemic manifestation of SLE. Dilatation of scleral vessels has significantly decreased and her vision remains unimpaired

CONCLUSION

Scleritis may present as an early or isolated manifestation of systemic immune-mediated disease. Therefore, a comprehensive examination is of the essence to exclude multisystem disease and treat the underlying cause. This should start with a thorough medical history with an extensive physical and laboratory examination, in addition to a full ophthalmology examination, even when there is no systemic manifestation.

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