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Case Report

ARCHIVES OF CLINICAL AND MEDICAL CASE REPORTS

ISSN: 2575-9655

Restoring Vision in SLE: Successful Management of Lupus Retinopathy – A Case Report

Md. Mehedi Hasan[1], Sanghita Banik Proma[2], Sakan Binte Imran[*3], Tanjila Hossain[4], Amiruzzaman[5]

Abstract

Background: Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease which significantly affects multiple organs, including the ocular structures. Lupus retinopathy is one of those ocular manifestations, characterized by retinal ischemia and microvascular damage.

Case Summary: During a study on retinal changes of Systemic Lupus Erythematosus (SLE) patients, we coincidently noticed some cases with lupus retinopathy, whose retinal changes were reversible after getting treatment for Systemic Lupus Erythematosus (SLE). This case report belongs to those cases. This case report presents a 38-year-old female who had initial complaints of blurred vision in right eye. The patient was concurrently diagnosed as Systemic lupus erythematosus (SLE), evidenced by polyarticular pain, fever and positive immunological and serological markers. The patient's visual symptoms significantly improved after receiving high dose glucocorticoids and immunosuppressive medications targeted towards her systemic symptoms of SLE. The follow-up exam showed that her vision had fully recovered and that his retinal lesions had resolved.

Conclusion: This case highlights the significance of lupus retinopathy as a potential early ocular manifestation of Systemic Lupus Erythematosus (SLE). Recognizing lupus retinopathy in its initial stages can not only prevent severe visual impairment but also facilitate the early diagnosis of SLE. Regular ophthalmologic screening is therefore essential in patients at risk, as it can play a pivotal role in the timely diagnosis and management of SLE.

Keywords: Systemic Lupus Erythematosus (SLE), Autoimmune disease, Lupus retinopathy, Ocular manifestations

Introduction

Systemic Lupus Erythematosus (SLE) is a connective tissue disorder of autoimmune origin which has a wide range of protean manifestations with variable course and prognosis [1]. It is a multi-system disorder that affects almost all organs, most commonly involving skin, joints and kidney [2]. Ocular involvement is reported in around one-third of the patients which can also present as an initial manifestation [2,3]. Lupus Retinopathy being the second most common ocular finding after keratoconjunctivitis sicca, is a sight-threatening condition which is characterized by deposition of immune complex in the retinal microvasculature leading to a chronological sequence of vascular occlusion, leakage, microinfarcts formation and retinal vasculitis

Affiliation:

[1]Postgraduate trainee, Department of Medicine, Sir Salimullah Medical College Mitford Hospital, Dhaka, Bangladesh

[2]Master's Student, University College London, London, England

[3]Medical Graduate, Sir Salimullah Medical College Mitford Hospital, Dhaka, Bangladesh

[4]Assistant Professor, Department of Ophthalmology, Sir Salimullah Medical College Mitford Hospital, Dhaka, Bangladesh

[5]Associate Professor, Department of Medicine, Sir Salimullah Medical College Mitford Hospital, Dhaka, Bangladesh

***Corresponding Author**
Sakan Binte Imran, Medical Graduate, Sir Salimullah Medical College Mitford Hospital, Dhaka, Bangladesh

Citation: Mehedi Hasan, Sanghita Banik Proma, Sakan Binte Imran, Tanjila Hossain, Amiruzzaman. Restoring Vision in SLE: Successful Management of Lupus Retinopathy – A Case Report. Archives of Clinical and Medical Case Reports. 9 (2025): 65-70.

Received: March 25, 2025
Accepted: April 03, 2025
Published: April 13, 2025

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DOI:10.26502/acmcr.96550712

[2]. Cotton wool exudates, hemorrhages, vascular tortuosity and attenuation, optic neuritis, optic atrophy, papilledema, macular edema, choroidopathy lastly chorioretinopathy are all possible manifestations of Lupus retinopathy, denoting that it has a wide range of clinical manifestations, from being asymptomatic to severe loss of vision [4]. We report a case of symptomatic lupus retinopathy with significant vision loss, whose ocular manifestation was reversed due to early recognition and a multidisciplinary treatment approach, both clinically and morphologically.

Case Presentation

A 38-year-old woman presented with a sudden onset of blurred vision in her right eye, experiencing no pain, itching, redness, or lacrimation. During ocular examination, visual acuity was noted as 6/60 in the right eye and 6/6 in the left eye.The patient couldn't recognize primary colours. No external ophthalmoplegia was present. Afterwards, Fundoscopy examination revealed that the media was clear, red reflex, and fundal glow were present in both eyes. The right (Rt) eye showed mild hyperemic disc with blurred nasal margin. There were widespread cotton wool exudates, notably on 5, 8, 10, and 11 O'clock positions of the right fundus (Figure 1). The left fundus showed no abnormality. Fundus fluorescein angiography (FFA) of Rt eye showing colour fundus and red-free photographs & successive phase of arteriolar- arteriovenous & venous phase which showed cotton wool spots present and temporal pallor disc (Figure 2). Optical Coherence Tomography (OCT) of the right eye revealed no abnormality (Figure 3).

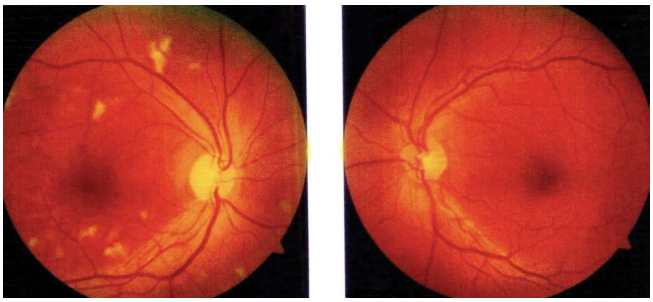

Figure 1: Fundal Photograph showing right-sided pale optic disc and cotton wool exudates. The left fundal was normal.

Upon querying, the patient revealed she had a feverish feeling, generalized body aches, and multiple joint pain with significant morning stiffness for the last 2 months. Initially involving the wrist, the joint pain gradually involved both large and small joints. She didn't have any known comorbidity like Diabetes Mellitus(DM), Hypertension (HTN) or hypercoagulable conditions. On general and systemic examination, body temperature was 100 degrees F, joint tenderness was present with a tenderness score of 2/4, and mild edema. There was no joint swelling or deformity. Routine investigations showed increased ESR, urine R/E

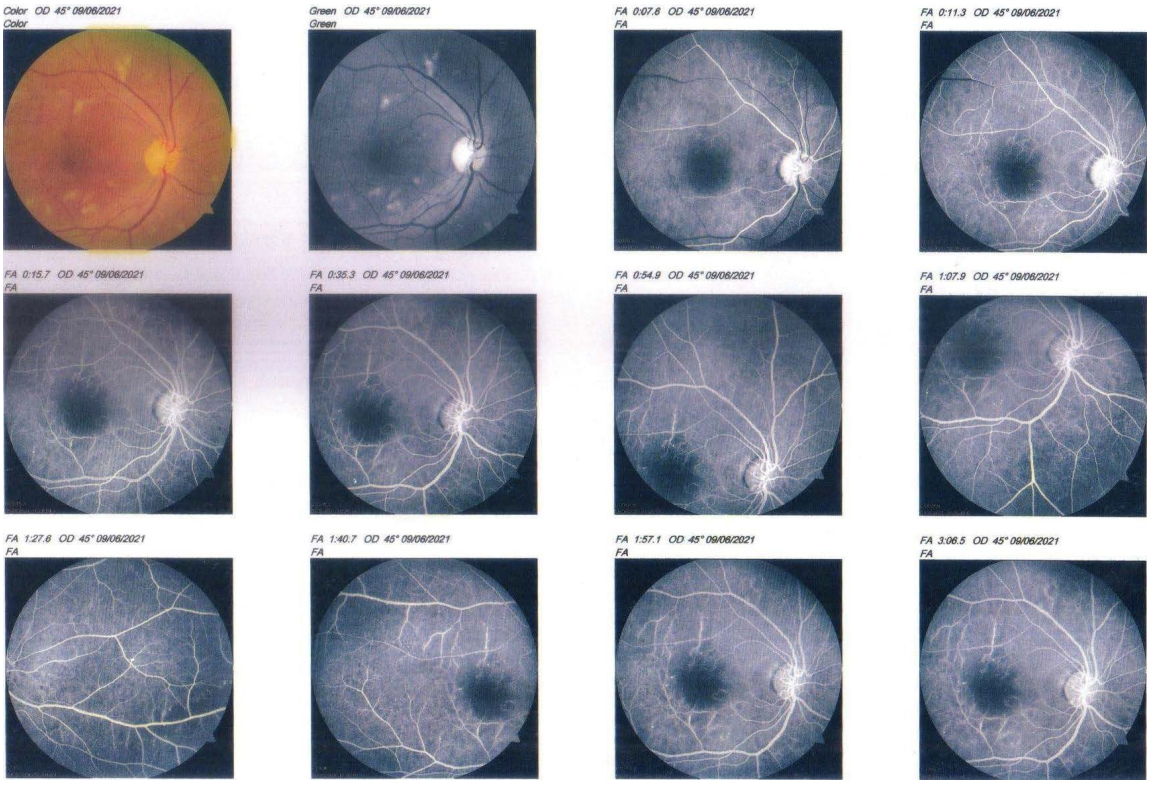

Figure 2: Fundus fluorescein angiography (FFA) of right eye showing colour fundus, red-free photographs & successive phase of arteriolar-arteriovenous & venous phase revealed cotton wool spots and pallor optic disc.

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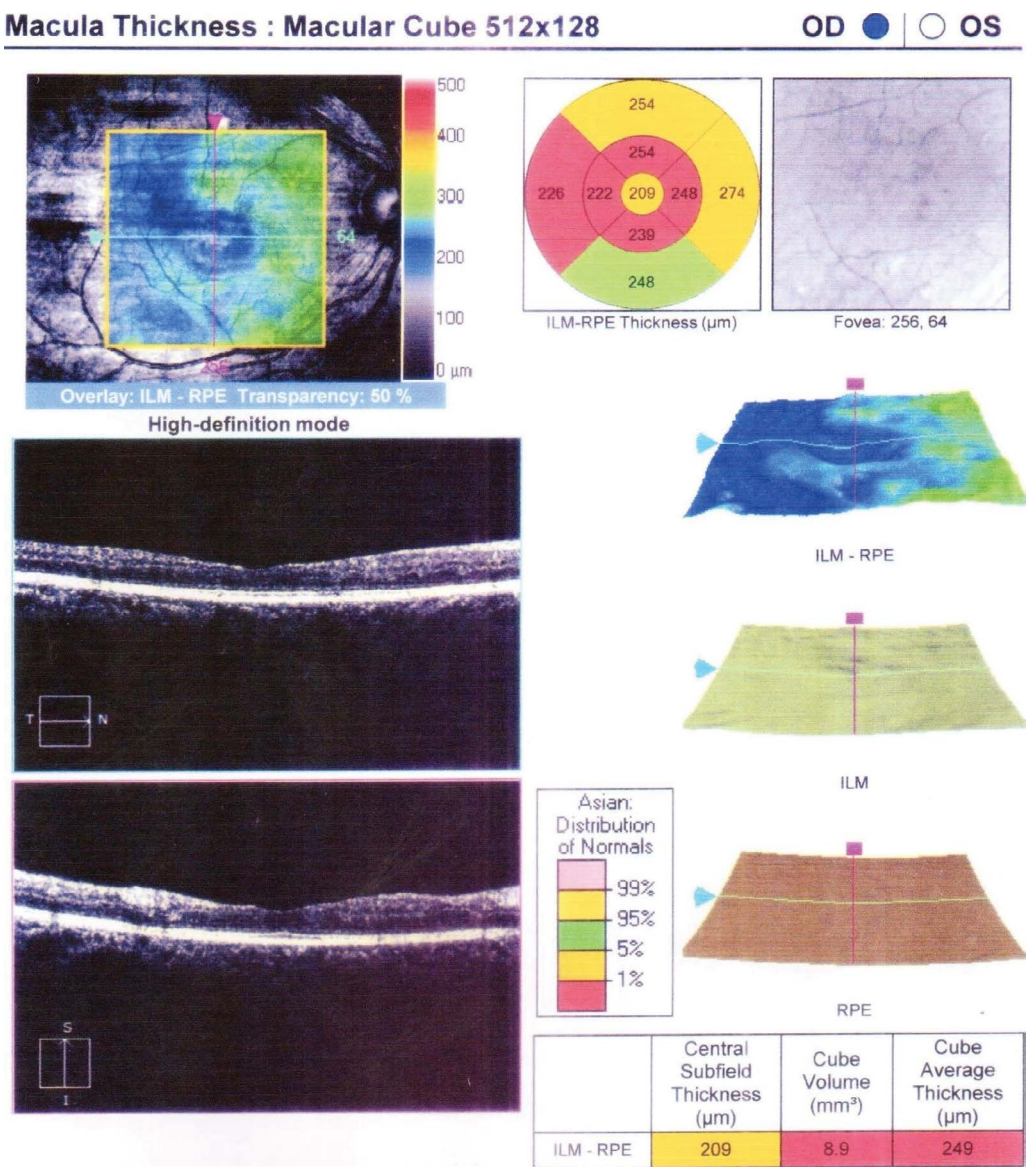

Figure 3: Optical Coherence Tomography (OCT) Macula of right eye revealed no abnormality of the macula.

shows 3+ proteinuria, and 24-hour Urinary Total Protein (UTP) is 2.4 gm (Table 1). Immunological investigation showed ANA was strongly positive, and Anti-dsDNA was positive. Significant consumption of complements C3 and C4 was also observed. So, the patient was ultimately diagnosed with a case of Systemic Lupus Erythematosus (SLE) according to the ACR diagnostic criteria with the following complications, lupus nephritis and lupus retinopathy. Her SLEDAI score was 23. Therefore, the patient was labeled as a case of very severe SLE. She was treated with a high dose of methylprednisolone followed by oral prednisolone and mycophenolate mofetil (MMF) targeting the systemic features of Systemic Lupus Erythematosus (SLE).

After treatment, the patient's systemic features were gradually improving. After 1 year of follow-up, her joint pain was better, having no fever or edema. Her 24-hours UTP became 0.2 gm. This time, her SLEDAI score was 4. On visual assessment, her vision was better with a visual acuity of 6/18. Fundoscopic examination revealed reduced optic disc paleness on the temporal side and there were no cotton wool exudates present (Figure 4). Thus, it became evident that her vision was partially improved with restored fundus morphology by providing treatment focusing on her systematic symptoms. Upon subsequent follow-up of the patient the fundoscopic finding of the eye showed retinopathy was reversed, yet the pale optic disc persisted to some extent but was better than before (Figure 5). Optical Coherence Tomography (OCT) of the right macula showed normal findings (Figure 6).

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Table 1: Investigation profile

Parameters	At the time of diagnosis	On follow-up
ANA	Strongly positive	Within normal limits
Anti-ds DNA	Positive	Within normal limit
C3, C4	Reduced	Within normal limit
Urine R/E	Protein +++	Protein(trace)
24 hours UTP	2.4 gm	0.2 gm (within normal limit)
SLEDAI	23 (very severe)	4 (mild)
Fundoscopic finding	Cotton wool exudates	Temporal pallor persist
(Right eye)	Optic disc hyperemic with blurred margin	(less than before)
Fundoscopic finding (Left eye)	Normal	Normal

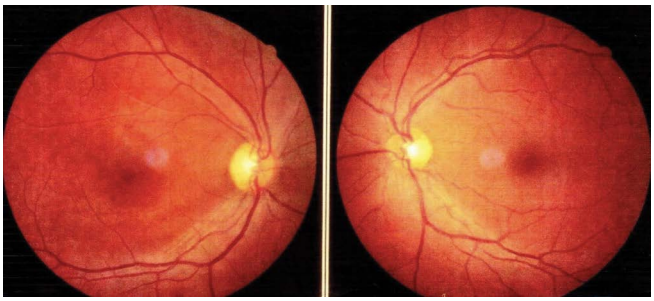

Figure 4: Fundal photograph showed reduced optic paleness on temporal side of the right eye

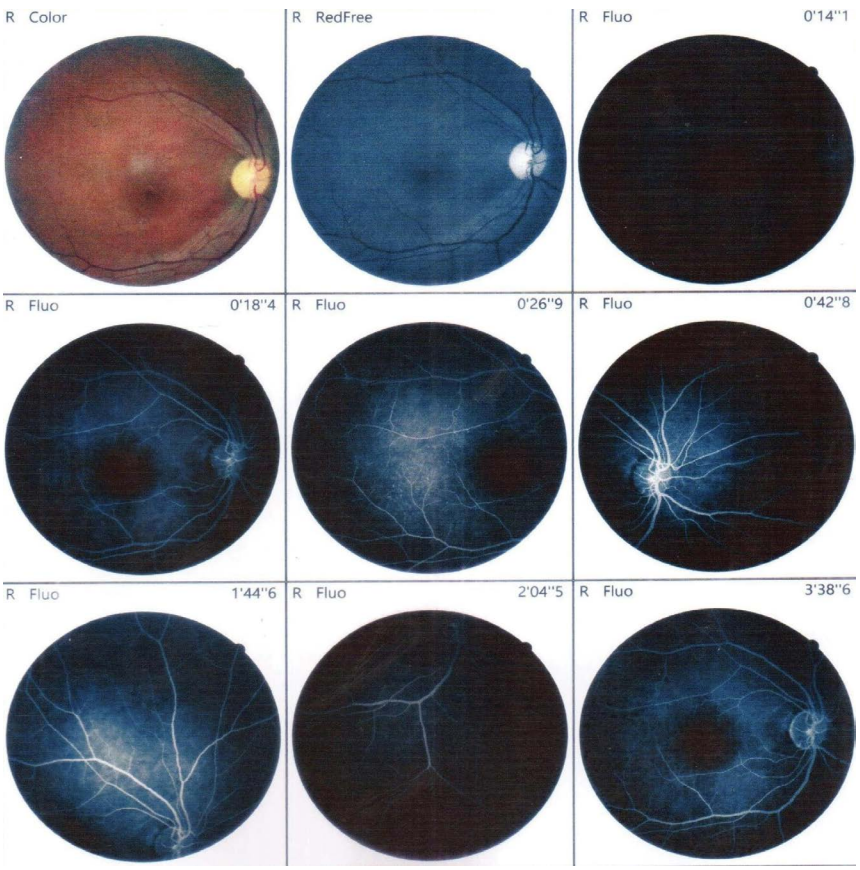

Figure 5: Fundus fluorescein angiography (FFA) of the right eye showing pale optic disc.

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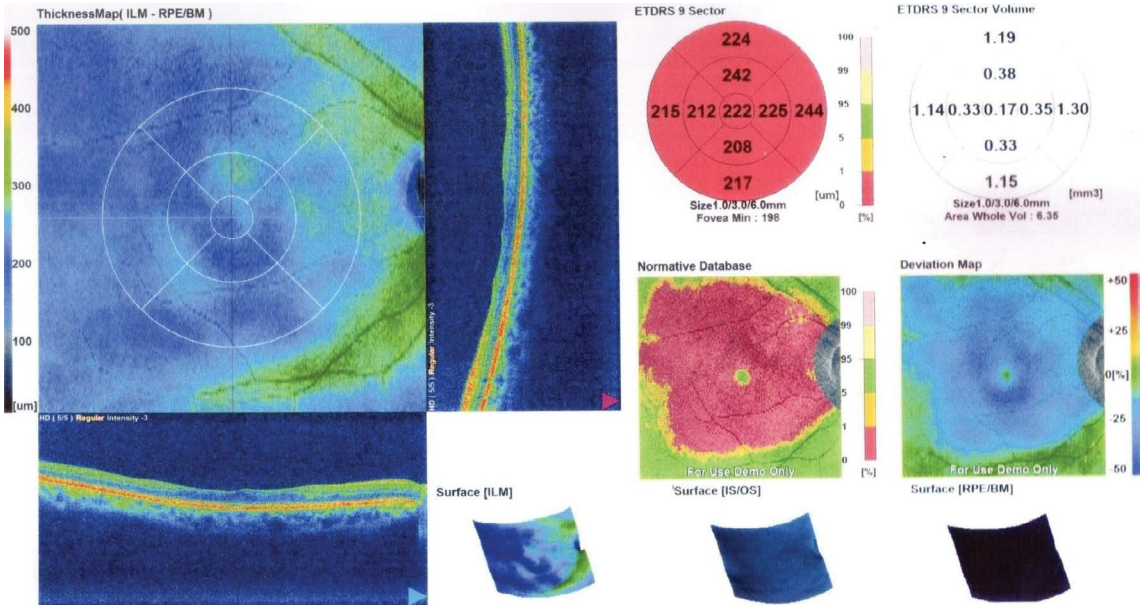

Figure 6: Fundus fluorescein Tomography (OCT) Macula showing normal findings.

Discussion

Lupus retinopathy is an important ocular manifestation, which occurs with an incidence of 3-29% [5]. The fate of SLE is worse with retinopathy compared to SLE without retinopathy as it serves as a marker of poor prognosis[4]. Even sudden and painless loss of vision as an ocular complication due to vaso-occlusive retinopathy over a short span of time has been reported [6]. Emphasizing the urgency of early detection followed by prompt management can thereby lead to a better prognosis of both systemic and retinal lupus [7]. Our patient is one of the living pieces of evidence of such an incident. The case presented here acts as a strong illustration, showing enhancement in both visual and morphological results of lupus retinopathy, even without retina-targeted treatments like laser photocoagulation or intravitreal anti-VEGF injections. Although ACR diagnostic criteria don't include Lupus retinopathy as a diagnostic tool, it can accurately indicate active systemic lupus activity which demands proper evaluation and appropriate management for better prognosis [8]. It accounts for further studies and concurrent research to support and establish their prior statement. As evidence backed up the fact that Lupus retinopathy is rather seen at an earlier stage of SLE and even makes the prognosis worse, it surely needs to be considered as an inclusion criteria going onwards. So, early diagnosis and adequate treatment of SLE can be ensured.

Conclusion

Systemic lupus erythematosus (SLE) is one of the numerous systemic disorders that have the fundus serving as a concealed mirror which reflects not only its progression but also its potential reversibility status. Therefore, fundoscopic findings can play an essential role as a diagnostic method, indicator of disease severity, and a prognostic element for SLE management.

Recommendation

Lupus retinopathy, frequently one of the initial signs of Systemic Lupus Erythematosus (SLE), underscores the significance of prompt diagnosis and intervention, rendering this condition treatable.Thus, the fundoscopic examination is strongly recommended as a standard screening method both at the time of diagnosis and during subsequent follow-up of SLE patients, as highlighted by this case report.

Declarations

Ethical Clearance

Complete information was given to the patient and written informed consent was obtained from the patient.

Consent to Publication

Written informed consent was obtained from the participant for publication of the case report.

Data and materials availability Declaration

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Competing Interest

The authors declare no competing interest.

Funding Declaration

The case report titled "Restoring Vision in SLE:

Md. Hasan M, et al., Arch Clin Med Case Rep 2025
DOI:10.26502/acmcr.96550712

Successful management of Lupus Retinopathy – A case Report" was conducted at Sir Salimullah Medical College Mitford Hospital. No funding was provided for this purpose.

Authors Contribution

Dr. Md Mehedi Hasan, Dr. Sanghita Banik Proma and Dr. Sakan Binte Imran : writing the original draft and revising.

Dr. Tanjila Hossain: validation and reviewing the draft.

Dr. Amiruzzaman: supervision, reviewing and revising the manuscript. All authors read and approved the final manuscript.

Acknowledgements: Not applicable

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