Brief Communication

# Retinal Vasculitis as Early Manifestation of Juvenile Systemic Lupus Erythematosus

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## ABSTRACT

Systemic lupus erythematosus (SLE) is a chronic, autoimmune disorder involving multiple organ systems with variability in its clinical features and complexity of disease course. Ocular manifestations occur in one-third of SLE patients. Vision-threatening retinal vasculitis may be the initial eye presentation in such cases. We herein present a case of a teenage girl who exhibits signs and symptoms of retinal vasculitis after the initial diagnosis of SLE.

**Key Words:** Autoimmune Diseases, Retinal Vasculitis, Systemic Lupus Erythematosus.

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## INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic disease with a clinical spectrum that ranges from cutaneous manifestations to various systemic involvements due to autoimmunity against self-nuclear antigens. Multiple aetiological factors including environment, viral and genetic susceptibility are postulated in the pathogenesis of SLE.[1]

Ocular changes can be detected in one-third of the patients with SLE.[2] SLE affects different parts of the eye, including eyelid, ocular adnexa, sclera, cornea, uvea, retina, and optic nerve. Secondary Sjogren syndrome is a commonly reported manifestation of SLE. Other manifestations include episcleritis, scleritis, iridocyclitis, retinopathy, choroidopathy, and optic neuropathy. Vision loss results from choroidal and retinal diseases.[3]

Retinal vasculitis is a vision-threatening condition of retina, in which, inflammation of the retinal blood vessels occurs due to infection or inflammatory processes, leading to disruption of the blood-retina barrier.[4]

We present a case of a 15-year-old SLE patient, who showed signs of retinal vasculitis in the early disease course.

## Case Presentation

A 15-year old female, presented to Dermatology Department, Sahiwal Teaching Hospital, Sahiwal with complaints of high-grade fever and headache. It was intermittent and had been present for 6 months. Malar rash appeared during the last 2 months. There was also history of photosensitivity, mouth ulceration, and loss of hair for 2 months. There were no joint pains, Raynaud's phenomenon, and other systemic complaints. On general physical examination, blood pressure was 100/70, pulseat 90/min, respiratory rate was 18/min, and temperature was 98°F. She was pale with no other findings of peripheral oedema or lymphadenopathy. Cutaneous examination revealed erythematous scaly plaques, involving nose and cheeks in butterfly distribution (Figure. 1). Scalp examination showed non-scaring patches of alopecia with lustreless dry hair (Figure 2). There was erythema and erosions involving gingival mucosa and hard palate. Dusky erythematous patches and papules were noticedon hands and feet, suggestive of vasculitic lesions. Laboratory investigations revealed haemoglobin, 8.9

g/dl, WBC 1.9 × $10^9$/L with lymphopenia (0.6 × $10^9$/l), platelet count 38 × $10^9$/l, and liver and renal function tests were normal. ANA was positive (1:512) and low complement levels C3 (32 mg/dl) and C4 (8 mg/dl). Serum Anti- dsDNA was negative. She was diagnosed with systemic lupus erythematosus fulfilling 5 clinical and 2 immunological criteria of the 2012 SLICC Criteria.

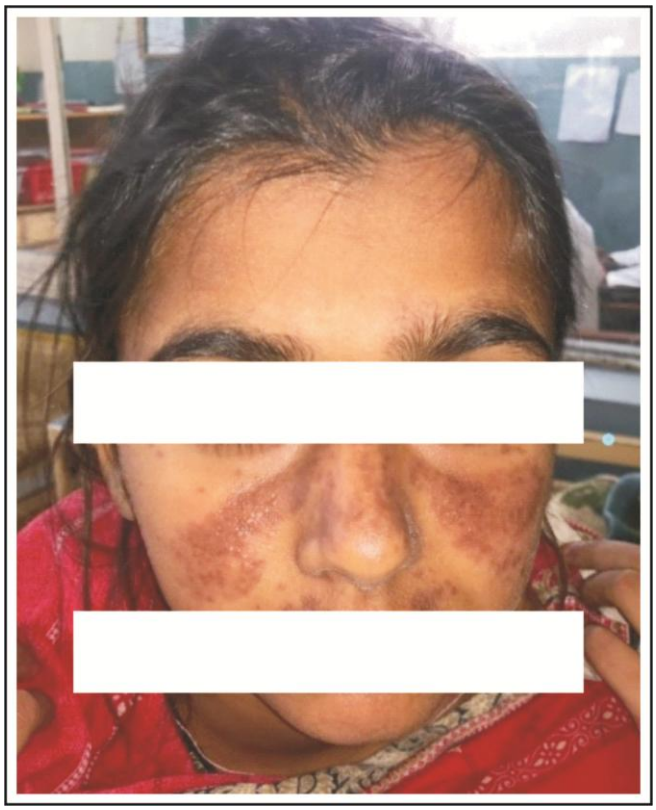

**Figure 1:** Malar Rash on face.

Dexamethasone 6mg (1.5 cc once daily), Hydroxychloroquine 200 mg twice daily, sunblock, and topical corticosteroids were started. Her cutaneous rash improved with topical medication, but her headache and fever were not settled. A lumbar puncture was performed to rule out CNS infection and inflammation, but CSF analysis revealed no findings. She was given Methyl Prednisolone sodium Succinate, 500 mg intravenous, 6 hourly for 3 days as a pulse therapy and her symptoms improved. She was discharged and called for a follow-up after 2 weeks.

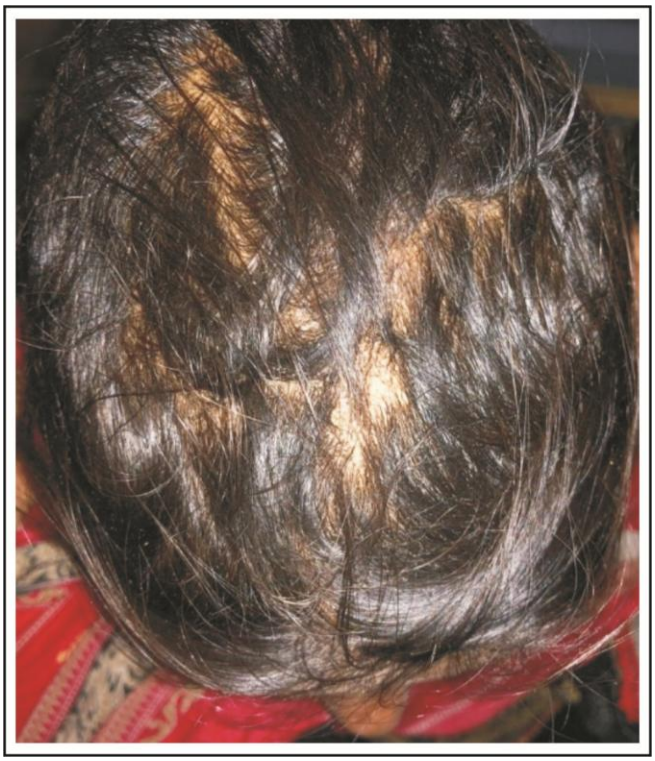

**Figure 2:** Non-scaring patches of alopecia with lustreless dry hair.

The patient presented in Dermatology after 15 days with complaints of painless blurring of vision in both eyes, associated with severe headache for 2 days. A complete blood count showed; WBC 4.9 × $10^9$/l, Hb 9.2 g/dl, PLT 245 × $10.^9$ Eye examination was done by the Consultant Ophthalmologist. Visual acuity was 3/60 in both eyes and the fundus examination revealed bilateral intraretinal and subretinal fluid, cotton wool spots, retinal vasculitis, and increased macular thickness (Figure 3). Fluorescein angiography showed bilateral decreased retinal vascularity, bilateral capillary closure, and decreased vascularity in the choroid. These features were suggestive of retinal vasculitis secondary to SLE. Prednisolone 40mg once daily was started with subsequent tapering by 5 mg of Prednisolone weekly. Azathioprine was added in the dose of 50 mg twice daily. The patient reported improvement in her visual acuity whichrecovered to 6/6 in the right eye and 6/9 in her left eye at 6 weeks follow up.

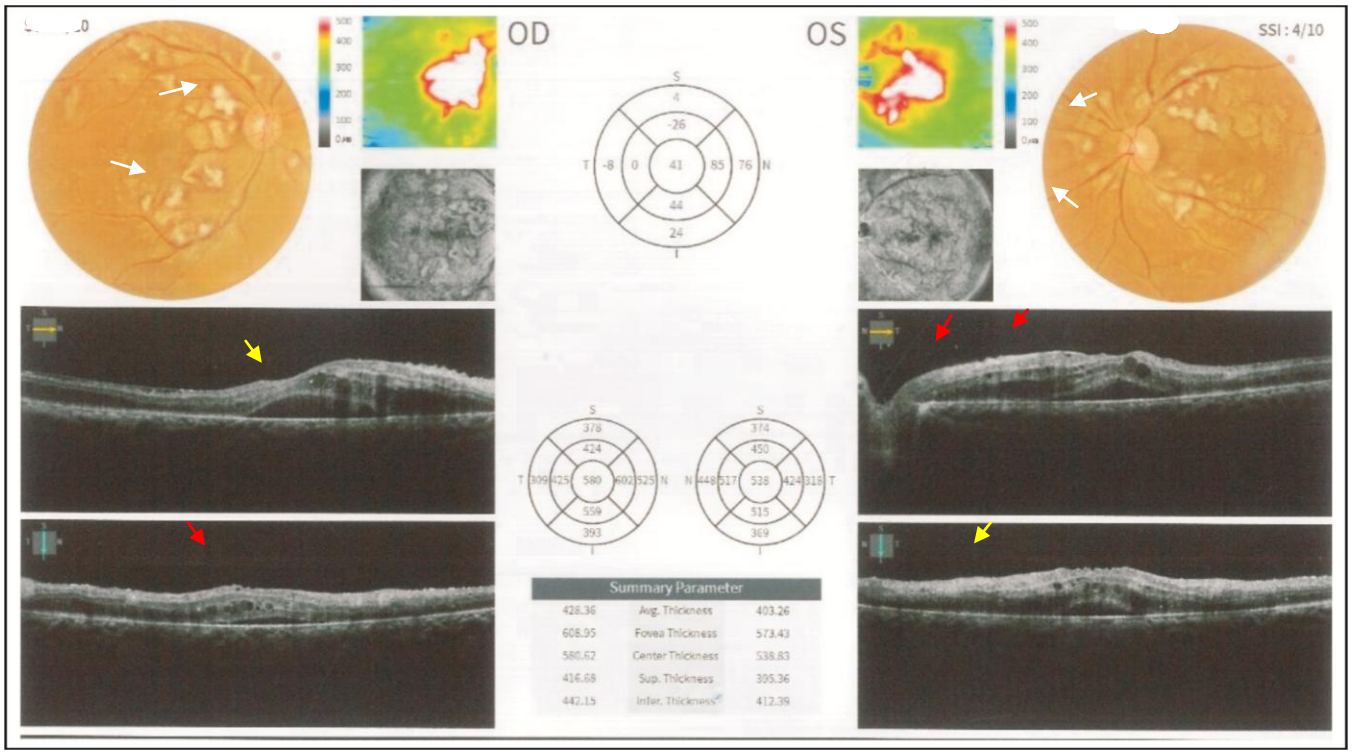


**Figure 3:** Fundus Picture and OCT scan. (Cotton wool spot with white arrows, Intra-retinal fluid with red arrows, Sub-retinal fluid with yellow arrows.

## DISCUSSION

SLE is an autoimmune disease with the potential to affect any organ. Every case of SLE must be examined by an ophthalmologist. Likewise, each case of retinal vasculitis must be ruled out for the presence of SLE. Among the spectrum of retinal involvement, microangiopathy is the mildest form. It usually presents in the form of cotton wool spots, retina haemorrhages, retinal oedema, and hard exudates. Good visual recovery generally follows.[5] Retinal vasculitis may present in the form of vascular sheathing that most commonly involves retinal arterioles, but retinal venules may be affected. Retinal vasculitis in the severe form that leads to vascular occlusion. Vascular occlusion may present as capillary dropout, central or branch retinal artery occlusion, retinal vein occlusion, and choroidal ischemia. After the vascular occlusive event, visual loss of varied severity may follow.[6] With prompt diagnosis and appropriate treatment, complications can be mitigated to preserve the useful vision. Although the severity of ocular involvement may or may not correlate with the severity of systemic involvement by SLE, it is mandatory to perform retinal examination in all cases of systemic vasculitis.[7,8] Conjunctiva and retina are the places where vasculature can be directly examined due to the presence of transparent overlying media.

In the present case, retinal involvement occurred following the systemic manifestation of SLE. Our findings are in accordance with the findings of the case presented by Noh and co-authors where the SLE case presented with peripheral retinal haemorrhages at routine check-up.[9] Roongta R et al reported a case of child who presentedwith visual loss as presenting compliant of SLE.[8] In our case, the patient suffered from a mild degree of vascular occlusion and vision deterioration. Timely diagnosis and prompt treatment resulted in visual recovery. Moreover, in young patients, retinal vascular walls have abundant elastic tissue. That factor along with good microvascular circulation could have led to visual recovery.[6] In contrast to our study, another report showed a severe visual loss in more than 50% of cases following SLE-associated retinal vascular occlusion despite treatment.[10]

SLE is a systemic disease that involves multiple organs not necessarily at the same time. A multidisciplinary approach is of utmost importance for the management of the disease. The present case points out the importance of early diagnosis of retinal

vasculitis and timely institution oftreatment resulting in good visual recovery. An ophthalmologist should be part of the SLE management team who can watch for signs of SLE and drug adverse reactions that can happen during the treatment.

## CONCLUSION

At the time of diagnosis of SLE as well as in follow up visits patient needs to be referred by Physician or Dermatologist to Ophthalmologist for comprehensive ophthalmic examination regardless of the presence of ophthalmic complaints.

**Conflict of Interest:** Authors declared no conflict of interest.

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## Authors' Designation and Contribution

Shahana Noor; Postgraduate Resident: *Concepts, Design, Manuscript preparation.*

Zahid Rafiq; Assistant Professor; *Literature search, Critical revision.*

Ahmad Zeeshan Jamil; Professor: *Manuscript writing, Proof reading and Final approval.*