

Conjunctival chemosis and isolated pericardial effusion as initial manifestations of systemic lupus erythematosus

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Abstract

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease with a wide spectrum of clinical manifestations, often making diagnosis challenging, particularly in atypical presentations. Ophthalmic involvement in SLE is relatively common; however, initial presentation with ocular symptoms is uncommon, and presentation with chemosis is exceedingly rare. The case of a 34-year-old previously healthy female who came to the emergency department of Shifa International Hospital, Islamabad in September 2022, is presented. She had bilateral chemosis and pericardial effusion and was diagnosed as SLE on the basis of strongly positive immunology. The patient demonstrated a favourable clinical response to corticosteroids and immunosuppressive therapy.

Keywords: systemic lupus erythematosus, chemosis, pericardial effusion, cyclophosphamide.

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Introduction

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune inflammatory disorder that can affect virtually any organ of the body. Ocular involvement typically occurs later in the disease course and is often associated with more severe systemic disease. Approximately one-third of patients with SLE exhibit ophthalmic signs and symptoms, which may range from mild to severe and potentially vision-threatening.¹

Disease manifestations may include involvement of the adnexa, orbit, retro-orbital tissue, ocular structures and the optic nerve. The most common ocular manifestation, keratoconjunctivitis sicca, usually results in only mild

irritation of the eye.²

This article focusses on chemosis, characterised by conjunctival oedema, as the initial presentation of SLE. Although not life threatening, chemosis can in turn lead to other ocular manifestations such as decreased visual acuity, increased susceptibility to developing bacterial keratitis and retro-orbital tissue involvement.

Case Report

A 34-year-old female presented to the emergency department of Shifa International Hospital, Islamabad in September 2022 with episodes of fainting spells and inability to tolerate oral intake due to nausea and vomiting of 3 days duration. She reported a one-year history of ill health, with significant deterioration over the preceding two weeks, during which she became largely bed-bound due to generalized weakness and reduced oral intake.

She had consulted multiple doctors in the previous month due to the development of facial, periorbital, and eyelid swelling. She was unable to achieve complete eyelid closure, resulting in blurred vision and a foreign body (gritty) sensation.

Further history revealed intermittent low-grade fever and arthralgias for the past year. She also reported mild Raynaud-like symptoms in winter for the past 4 years, for

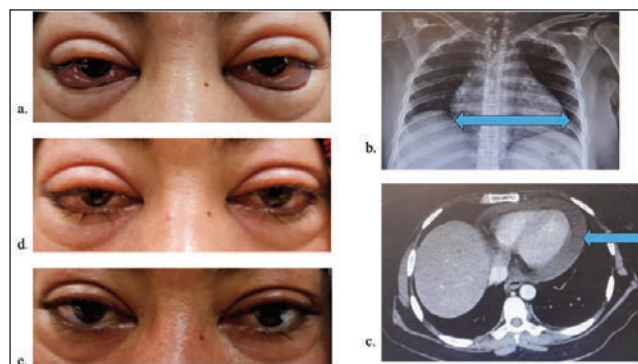


Figure-1: a) Initial presentation showing severe conjunctival swelling and hyperaemia, and swollen lids bilaterally. b) Chest X-ray showing increased width of the cardiac shadow. c) CT scan with contrast showing moderate pericardial effusion. d) During treatment, resolving conjunctival and eyelids' swelling. e) Resolved conjunctival oedema post treatment.

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Table 1A and 1B: Baseline workup and Immunological workup.

Investigation	Patient's result	Reference range
Table 1A – Baseline Workup		
Complete Blood Count		
White blood cells, Total	7420	4000 - 11000 /UL
Haemoglobin	9.1	11.6 - 16.5 g/dL
Haematocrit	28.7	38 - 47%
MCV	64.5	80 - 90 fL
Platelets	129,000	150,000 - 400,000 /UL
Red blood cells, Total	4.45	3.8 - 5.8 m/UL
RDW	21.7	11.5 - 13.6 %
Inflammatory Markers		
CRP	16.58	< 5 mg/L
ESR	31	6 - 20 mm fall in the first hour
Procalcitonin	0.17	< 0.1 ng/ml
Electrolytes and Renal Function Tests		
Serum Sodium	135	136 - 145 mEq/L
Serum Potassium	3.6	3.5 - 5.1 mEq/L
Serum Bicarbonate	21	22 - 29 mEq/L
Serum Urea	23.54	16.6 - 48.5 mg/dl
Serum Creatinine	0.73	0.57 - 1.11 mg/dl
Estimated GFR	97.66	> 60 ml/min/1.73m ²
Liver Function Tests		
ALT	54	< 35 U/L
AST	60	< 35 U/L
ALP	112	40-130 U/L
Total Bilirubin	0.34	< 1.2 mg/dl
Direct Bilirubin	0.12	< 0.30 mg/dl
Serum Albumin	2.9	3.9 - 5 mg/dl
Cardiac Markers		
CPK	27	26 - 192 U/L
CKMB	0.4	< 3.4 ng/ml
Troponin I	4.3	< 15.6 pg/ml
BNP	41.9	< 100 pg/ml
Endocrine Workup		
TSH	1.76	0.4 - 4.2 uIU/ml
Free T4	1.24	0.70 - 1.70 ng/dl
T3	86	80 - 200 ng/dl
HbA1c	5.3	< 5.7 %
COVID-19/SARSCOV-2 by PCR	Negative	
Table 1B – Immunological Workup		
ANA	3+ speckled (titre > 3200)	Negative
Nucleosome antibodies	18	Negative < 5 Equivocal 5-10 Positive > 10
dsDNA antibodies	26	
Anti-Histone antibodies	< 0.1	
Anti-Sm antibodies	61	
Anti-RNP antibodies	87	
AntiSm/Anti-RNP antibodies	100	
Anti-SSA (Ro), Anti-SSb (La), Anti Jo-1, Anti Scl-70	0.1	
Complement levels		
C3	0.33	0.9 - 1.80 g/dL
C4	0.02	0.10 - 0.40 g/dL

MCV - Mean Corpuscular Volume, RDW - Red Cell Distribution Width, CRP - C Reactive Protein, ESR - Erythrocyte Sedimentation Rate, GFR - Glomerular Filtration Rate, ALT - Alanine Transaminase, AST - Aspartate Transaminase, ALP - Alkaline Phosphatase, CPK - Creatine Phosphokinase, CKMB - Creatine Kinase MB, BNP - Brain Natriuretic Peptide, TSH - Thyroid Stimulating Hormone, T4 - Thyroxine, T3 - Tri-iodothyronine.

ANA - anti-nuclear antibody, dsDNA - double-stranded deoxyribonucleic acid, anti-Sm - anti-Smith, anti-RNP - anti-ribonucleoprotein.

which she had not sought medical attention. There was no history of malar rash, photosensitivity, orogenital ulcers or hair loss. Her past surgical history was unremarkable, and her family history was significant for rheumatoid arthritis in her mother. She had 2 children and no history of miscarriages.

On examination, the patient was alert but markedly weak requiring a wheelchair for mobility. Her pulse rate was 100/min, and she had a blood pressure of 122/85mmHg. She appeared slightly pale with marked facial puffiness. There was significant bilateral conjunctival hyperaemia and swelling, suggestive of chemosis, with associated eyelid swelling; she was unable to fully close her eyelids. (Figure 1a). She had slightly puffy fingers, but no clear-cut synovitis, and hand grip was normal. There was mild ankle oedema. Chest sounds were normal on auscultation.

Following admission to the medical department, a thorough initial workup was undertaken (Table 1A) which revealed raised inflammatory markers, microcytic anaemia and thrombocytopenia. Renal function was normal, while liver function tests showed transaminitis and hypoalbuminaemia. Urine showed trace protein.

ECG displayed mild tachycardia (108/minute), normal rhythm with no acute changes. Cardiac markers were normal. Chest X-ray showed an increased cardiac shadow with clear lung fields. (Figure 1b).

Given the facial puffiness, CT chest, abdomen and pelvis with contrast was performed to exclude SVC syndrome and malignancy, and showed only moderate pericardial effusion. (Figure 1c). Echocardiography later confirmed a moderate global pericardial effusion measuring 1.7cm with no evidence of tamponade or any structural abnormalities, and an ejection fraction of 60%.

Thyroid-associated orbitopathy was initially suspected; however, this was not supported as thyroid function tests were within normal limits, despite elevated antithyroglobulin antibodies.

A previously performed magnetic resonance imaging (MRI) scan of the orbits showed no evidence of a retro-orbital mass or other significant pathology.

ANA was strongly positive which prompted detailed immunologic workup (Table 1B); this revealed strongly positive anti-dsDNA, anti-Sm, anti-nucleosome and anti-RNP antibodies. Complement levels were low. Antiphospholipid antibody screen, Rheumatoid Factor and Antineutrophil cytoplasmic antibodies were negative.

The patient fulfilled the latest EULAR/ACR 2019 criteria for SLE hence the diagnosis was made.

Prior to admission, she had been receiving low-dose oral prednisolone and moxifloxacin ophthalmic drops for one week, with minimal clinical improvement. The ophthalmology team was consulted; they placed her on steroid eye drops, and added artificial tears. Nausea and vomiting were symptomatically managed. A swallowing assessment was done by a speech therapist who advised initiating liquids and soft diet. Given the severity of symptoms, she was started on intravenous methylprednisolone 500mg daily. Improvement was seen in the next 48 hours, but due to personal reasons, she left against medical advice.

She followed up in the rheumatology clinic a few days later, feeling much better with reduced conjunctival hyperaemia and oedema. She was started on high-dose oral prednisolone 60mg daily and was counselled for stronger immunosuppression in the form of intravenous cyclophosphamide, to which she consented. She was given intravenous iron for microcytic anaemia after excluding thalassaemia trait. She received six cycles of intravenous cyclophosphamide at 2- to 3-week intervals, as per her availability, with a dose of 750mg for the first two cycles, and 500mg for the remaining four. Steroids were gradually tapered to 5mg daily over the following months.

She continued to improve, and chemosis resolved after the third cycle, allowing her to fully close her eyes. (Figure 1d).

Upon completion of 6 cycles, she was started on oral azathioprine as steroid-sparing therapy. After 2 months, she presented with pain in small joints and early morning stiffness. Examination revealed low-grade synovitis in hands with reduced grip strength. Therapy was subsequently changed to subcutaneous methotrexate, resulting in resolution of her arthritis.

She has been on follow-up and has remained largely symptom-free with steroids and immunomodulators. (Figure 1e).

Discussion

Chemosis as an isolated eye feature is a rare initial presenting complaint in patients with SLE. In our case, the patient's chief complaint was profound bilateral eye swelling and inability to close the eyes due to conjunctival oedema. Thyroid-associated orbitopathy was excluded, and a diagnosis of SLE was subsequently established.

The first case of chemosis in SLE was reported in 1982 by Zaini et al from Malaysia. They reported bilateral chemosis in a 21-year-old female who also had class II glomerulonephritis and was subsequently diagnosed with SLE.³ In 1992, chemosis and anaemia were the presenting features of SLE in a 36-year-old female.⁴

A case presented in 2010 described a 35-year-old female with a history of psoriasis who presented with chemosis along with pressured speech and suicidal thoughts, and was subsequently diagnosed with SLE.⁵ The most recently reported case published in 2022 is of a 29-year-old male who presented with severe bilateral chemosis and lupus psychosis, and was also found to have pleuritis, nephritis, and cerebral infarcts; he was diagnosed with SLE and antiphospholipid syndrome.⁶ As demonstrated, psychiatric symptoms were a common feature in previous cases, however, we found no such symptoms in our patient.

Only a few similar cases were found in our literature search, with third spacing in the pleural, pericardial, and peritoneal cavities, and pedal oedema, but none with an isolated pericardial effusion, as in our case.⁶⁻⁸

Two principal mechanisms for chemosis have been proposed in the reviewed case reports. One is based on an inflammatory cause, involving immune complex deposition,^{3,5} which is likely to be the mechanism in our case as well. Another possible theory is the role of hypoalbuminaemia^{3,7-9} with lupus nephritis. Although our patient had hypoalbuminaemia, there was no significant renal involvement, and only one similar case has been reported in the literature.⁴

All cases were initially treated with high dose pulse steroids, followed by maintenance steroids and immunosuppressive therapy which led to the resolution of chemosis. In many cases, cyclophosphamide is used.

To the best of our knowledge, no similar case has been reported from our region; therefore, this represents the first reported case from Pakistan and South Asia of SLE presenting with chemosis and isolated pericardial effusion.

Conclusion

In patients presenting with ocular signs such as chemosis, systemic lupus erythematosus (SLE) should be considered in the differential diagnosis, and an appropriate autoimmune workup should be undertaken. Clinicians should also evaluate for evidence of systemic involvement, which appears to be more common in patients presenting with chemosis.

This case highlights the importance of recognizing atypical ocular presentation of SLE, as prompt diagnosis and timely initiation of immunosuppressive therapy can lead to favourable clinical outcomes.

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TMH: Concept, critical review, editing, drafting, supervision and final approval.

ZM, MAC, MS & MAA: Data acquisition, analysis, writing, reviewing, editing, drafting and final approval.

MAA: Critical review, editing and final approval.