

Esophageal Dysmotility Disorder and Dysphagia as Initial Manifestations of Lupus

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Abstract

Systemic lupus erythematosus (SLE) is a chronic, multisystemic autoimmune disease that is diagnosed using specific clinical and biochemical inclusion criteria. Here, we report the case of a young adult who was initially diagnosed with dysphagia and later diagnosed with SLE. Other rheumatological and non-rheumatological conditions were ruled out, and serology revealed that SLE was the cause of the patient's symptoms.

To the best of our knowledge, no previous case studies or prior research identify dysphagia as a presenting symptom of SLE. While esophageal dysmotility ranging from ineffective esophageal motility (IEM) to achalasia can manifest as a later clinical consequence of SLE, it is rarely the first symptom of the disease. After diagnosis, the patient was treated with hydroxychloroquine and prednisolone, which led to significant improvement.

Categories: Internal Medicine, Rheumatology

Keywords: autoimmune disease, manometry, motility disorders, dysphagia, systemic lupus erythematosus

Introduction

Systemic lupus erythematosus (SLE) is an inflammatory autoimmune disease that affects various body organs and may present with a wide range of early clinical symptoms and signs [1]. According to the European League Against Rheumatism/American College of Rheumatology (EULAR/ACR), gastrointestinal symptoms are not one of the clinical inclusion criteria of SLE. However, mouth ulcers are listed with a score of 2 under mucocutaneous clinical criteria [1].

When analyzing the published literature, it was found that a few case reports mentioned uncommon gastrointestinal tract involvement as the first sign of SLE in a variety of conditions, including peritonitis, acute acalculous cholecystitis, abdominal cocoon, and protein-losing enteropathy.

In our case, a 35-year-old woman with no history of comorbid illness or medication use presented to the emergency department (ED). Her only symptom was difficulty in swallowing.

Case Presentation

A single, nonsmoking, 35-year-old Saudi female presented to the ED. Her chief complaint was difficulty in swallowing. Her symptoms began three months before. She first experienced difficulty swallowing liquids, which later extended to solids as well. The condition was progressive, and her symptoms were alleviated by fasting. Furthermore, the presentation was characterized by subjective fever, a weight loss of 9 kg, pain while swallowing, recurrent oral ulcers, hair loss, and joint pain, primarily affecting the shoulders and knees.

The patient reported symptoms occurring one week before the presentation. The results of an outpatient esophagogastroduodenoscopy (EGD) were also unremarkable.

Otherwise, the patient denied similar episodes before and did not report a history of any genital ulcers, morning stiffness, skin changes, or other joint involvement. A systematic review of medical, surgical, family history, medication/herbal, or cultural methods history was unremarkable.

The patient appeared underweight; her body mass index (BMI) was 14. She was hemodynamically stable and had no lymphadenopathy or swelling in the neck. No skin changes or specific patterns of hair loss were observed, and a systematic examination was otherwise unremarkable.

The initial laboratory findings, including inflammatory markers, were mostly normal (Appendix). She presented with bicytopenia, displaying low hemoglobin and low WBC count; a Coombs test was positive

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(Table 1).

Test	Result	Normal level
White blood cells (L ⁻¹)	3.4 × 10 ⁹	4-10
Platelet (L ⁻¹)	266 × 10 ⁹	150-400
Hemoglobin (g/dL)	8.9	12-15
Creatinine (mmol/L)	27	44-80
24-hour urine protein	0.2064	
ANA	1/1,280	Negative
Anti-DNA AB	Negative	Negative
C3 (g/L)	0.33	0.83-1.93
C4 (g/L)	0.078	0.15-0.57
B2-Glycoprotein 1 IgG	Negative	Negative
B2-Glycoprotein 1 IgM	Negative	Negative
Anti-cardiolipin IgG	Negative	Negative
Protease 3 antibodies	Negative	Negative
Fibrinogen (g/dL)	3.7	2-4
MCV (fL)	82.7	79-97
MCH (pg)	25.7	27-32
Anti-Smith	Negative	Negative <0.9
Anti-RO (SSA)	Positive	Negative <0.9
Anti-SSB-La	Negative	Negative <0.9
RF	<7 IU/mL	Negative
C-reactive protein	3.53 mg/L	0-4
Direct Coombs test	Positive	Negative
ESR (mm/hour)	13	0-20
Anti-Scl-70	Negative	Negative
Anti-Jo-1	Negative	Negative <0.9
Anti-centromere B	Negative	Negative <0.9

TABLE 1: Laboratory results of a 35-year-old female presented with difficulty swallowing.

Ig, immunoglobulin; ANA, antinuclear antibody; Anti-DNAAB, anti-DNA antibody; MCV, mean corpuscular volume; MCH, mean corpuscular hemoglobin

During admission, the patient underwent a barium swallow test, which revealed delayed gastric emptying (Figure 1). Her upper and lower scopes were normal (Figure 2), and her biopsies were negative. An eating disorder was excluded via a complete psychiatric evaluation. Malignancy was also excluded by negative pan-CT scans. Infection was ruled out due to unremarkable septic screens and cultures.

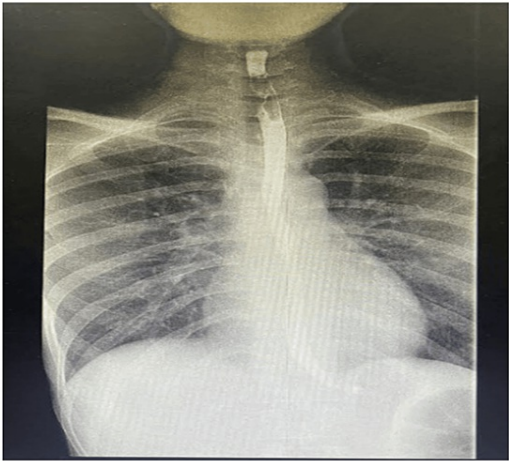


FIGURE 1: A case report with a barium swallow.

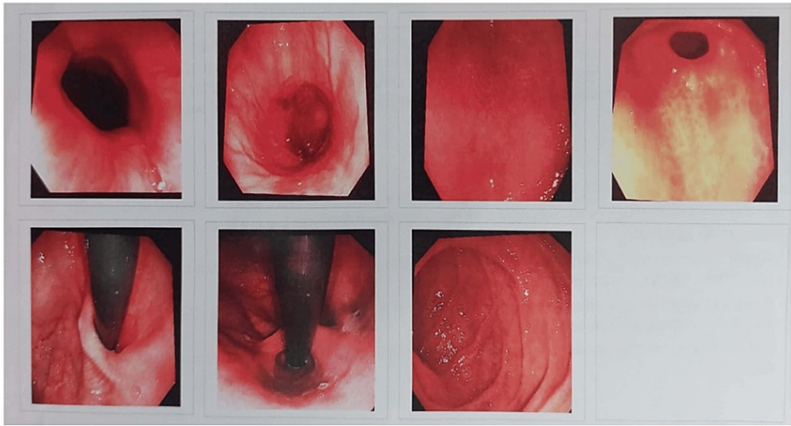


FIGURE 2: A case report with colonoscopy.

The patient’s positive Coombs test and delayed gastric emptying in barium swallow suggested an autoimmune disease, which seemed likely due to the patient’s age and rheumatological history.

The patient’s serology workup was positive for antinuclear antibody (ANA) and anti- rRNP and showed low C3 and C4 levels. She was negative for rheumatoid factor (RF), anti-Smith, and anti-cyclic citrullinated peptide (anti-CCP). The patient was thus diagnosed with lupus. According to the European League Against Rheumatism (EULAR) classification criteria, lupus is indicated by the presence of antibody markers, particularly when these markers appear in two organ systems. Our patient presented with three clinical criteria for lupus: joint involvement, oral ulcers, and non-scarring alopecia. Additionally, there were indications of leukopenia and autoimmune hemolysis. She also presented with two immunological criteria, resulting in a total score of 21 (Table 1).

Treatment was started immediately; the patient was administered 25 mg of steroids twice daily, 200 mg of hydroxychloroquine daily, and 5,000 IU of vitamin D with calcium weekly. She was also given proton pump inhibitors to alleviate her symptoms; manometry studies were conducted on an outpatient basis.

The patient’s clinical condition and eating improved, leading to weight gain and improved general health. She was discharged and instructed to follow up with the rheumatology and swallowing department. At a two-week follow-up, the patient’s dysphagia and odynophagia had improved; she had also gained 3 kg.

Discussion

SLE has a wide range of uncommon presentations reported in the literature due to its multi-organ involvement of unknown etiology [1], the European League Against Rheumatism (EULAR)/American College of Rheumatology (ACR) diagnostic criteria for SLE do not include gastrointestinal signs, apart from oral ulcers, which may be observed during the clinical examination [1].

A prospective study in the United States conducted from 2018 to 2020 used high-resolution manometry (HRM) to examine 1,003 patients with this rheumatological condition. Of these, 9% had gastrointestinal involvement, 2.7% had systemic sclerosis, 2% had rheumatoid arthritis, and 1% had SLE. Furthermore, 61% had esophageal motility disorders. Three categories of these disorders were most prevalent: 28% of patients had absent contractility, 21% had inefficient esophageal motility, and 12% had esophagogastric outflow blockage. This study also found that individuals with esophageal problems had larger upper esophageal sphincters, lower distal contractile integrals, poorer bolus clearance, and more hiatal hernias than healthy controls [2].

In a multicenter cohort study conducted in 2018, 389 patients who presented with early manifestations of SLE were compared to 227 patients who presented with SLE mimickers. The study's results showed a significant correlation ($P > 0.001$) between dysphagia and early SLE manifestation; however, the study did not state dysphagia as the main presenting symptom [3].

The gastrointestinal tract is involved in 40% of SLE patients. Most of these cases are linked to vasculitis activity, autoimmune tissue damage, complications of organ dysfunction, thromboembolic symptoms of antiphospholipid antibody syndrome, infections, decreased saliva production, or adverse drug reactions due to long-term use [4,5]. Moreover, 20% to 70% of dysphagia-related complaints are linked to heartburn, chest pain, odynophagia, or regurgitation with a motility disorder [5].

The history of esophageal abnormalities has been extensively researched in previous literature [6]. Sultan et al. were the first to report that the number of motility diseases was increasing [7]. Castrucci et al., Lapadula et al., and a Mexican study found hypokinetic processes mostly affecting the upper third of the esophagus, with a normal lower esophageal sphincter, in three distinct types of manometry studies [8,9,10]. The pathophysiological mechanism for SLE-related esophageal dysmotility has been suggested to be either related to Auerbach plexus nerves or smooth muscle vasculitis [11].

Previous case studies report different manifestations of motility disorders in patients with SLE. These include esophageal epidermolysis bullosa acquisita, vagal nerve dysfunction caused by a focal CNS lesion in the vagus nerve nuclei (leading to the need for a temporary gastric feeding tube), and a link between dysphagia and chest pain brought on by esophageal spasms [11]. To improve assessment and ensure appropriate treatment, patients first undergo a barium swallow evaluation before they are subjected to further manometry. When a motility disorder is suspected, the patient might undergo an endoscopy with an additional tissue biopsy to check for vasculitis, *Helicobacter pylori* infection, or ulceration [11].

Unfortunately, due to a dearth of evidence-based data on responses to treatment in SLE patients and since immunosuppressant medications may also initiate symptoms by predisposing patients to opportunistic infections, motility disorders in SLE patients are still not well understood. However, other diagnostic studies may help identify a specific etiology, which could support management. PPI or metoclopramide along with lifestyle and diet modifications can be effective initial therapies, as many SLE patients experience gastroesophageal reflux disease (GERD) [4,12]. Nitrates and calcium channel blockers can be used to treat esophageal spasms, and steroids may be used to treat SLE vasculitis. However, no research has yet aimed to determine the best immunosuppressant agent for patients with SLE [12].

Conclusions

Esophageal involvement is known to occur in some SLE patients over the course of the disease. Despite its rarity, SLE should be included in the differential diagnosis for any patient who exhibits the first predominant clinical indication of esophageal problems. Future research should explore the various types of dysphagia in more detail and measure the effectiveness of available treatments.

Appendices

Lab Location	Analysis	Result	Range From
Biochemistry	HCT	28.3	36 46
Hematology	MCV	81.1	79 97
Hematology	MCH	25.2	27 32
Biochemistry	MCHC	31.1	31.5 34.5
Hematology	RDW	24.2	11.6 14
Immunology -RRLBE	MPV	12.6	7.4 10.4
Hematology	PLI	391	150 400
Biochemistry	NRBC	0	
Hematology	NEU%	51.9	40 80
Serology KSMC	LYM%	33.5	20 40
Biochemistry	MON%	12.3	2 10
Hematology	EOS%	1.7	1 6
Blood Bank -RRLBB	BAS%	0.6	0 2
Biochemistry	NRBC%	0	
Hematology	PT sec	17.4	12.3 15.7
Biochemistry	PT INR	1.27	.8 1.2
Hematology	PTT Sec	37.4	26 45

a

Lab Location	Analysis	Result	Range From
Biochemistry	LYM	0.6	1 3
Hematology	MONO	0.22	.2 1
Hematology	EOS	0.03	.02 5
Biochemistry	BASO	0.01	.02 1
Hematology	Neutro	0.93	2 7
Immunology -RRLBE	WBC	1.79	4 10
Hematology	RBC	3.49	4.4 5.4
Biochemistry	HGB	8.8	12 15
Hematology	HCT	28.3	36 46
Serology KSMC	MCV	81.1	79 97
Biochemistry	MCH	25.2	27 32
Hematology	MCHC	31.1	31.5 34.5
Blood Bank -RRLBB	RDW	24.2	11.6 14
Biochemistry	MPV	12.6	7.4 10.4
Hematology	PLI	391	150 400
Biochemistry	NRBC	0	
Hematology	NEU%	51.9	40 80

b

FIGURE 3: (a) and (b) Test reports of hematology laboratory tests.

Lab Location	Analysis	Result	Range From
Biochemistry	ALT	19.7	33
Hematology	Alkaline Phosphatase	65	35 129
Hematology	AST	28.6	32
Biochemistry	Bilirubin - Direct	2	0 5
Hematology	Chloride Serum	107	98 107
Immunology -RRLBE	Creatinine Serum	29	44 80
Hematology	Glucose Random	3.98	4.11 5.89
Biochemistry	Potassium Serum	3.16	3.5 5.1
Hematology	Sodium Serum	140	136 145
Serology KSMC	CO2	27.2	22 31
Biochemistry	UREA (Bun)	0.9	2.14 7.14
Hematology	Bilirubin -Total	3.6	0 21
Blood Bank -RRLBB			
Biochemistry			
Hematology			
Biochemistry			
Hematology			

FIGURE 4: Test report of biochemistry laboratory findings.

Additional Information

Disclosures

Human subjects: Consent was obtained or waived by all participants in this study. King Saud Medical City issued approval King Saud Medical City Committee. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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