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Case Report

Gastrointestinal Manifestation of Systemic Lupus Erythematosus - Lupus Enteritis

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Abstract

Systemic Lupus Erythematosus (SLE) is a chronic inflammatory disorder with a heterogenous mode of presentation and clinical course. We present a case of a young female with an atypical presentation of SLE. A young woman with no known prior comorbidities presenting with the history of chronic diarrhea, vomiting and weight loss, followed by generalized weakness. Extensive workup was done and she was diagnosed with lupus enteritis based on history, laboratory investigations and findings on imaging. She was given pulse therapy to which she responded very well. The case describes the importance of high clinical suspicion and timely intervention as well as adds to the evidence on a rare clinical entity-lupus enteritis.

Keywords: SLE, Lupus enteritis, autoimmune disorders, chronic diarrhea.

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Introduction

Systemic Lupus Erythematosus (SLE) is a fairly common multi-system autoimmune disorder involving a wide spectrum of clinical features. The most frequent presenting symptoms are musculoskeletal or cutaneous involvement, followed by hematological, renal, cardiac, nervous system and gastrointestinal tract (GIT).[1] Gastrointestinal symptoms in SLE were first notified in 1895 and these may range from vomiting, abdominal pain to diarrhea.[2] The magnitude of GIT involvement is also greatly variable, ranging from hepatitis, pancreatitis, lupus enteritis to mesenteric vasculitis.[3] Lupus Enteritis (LE) was defined by the British Isles Lupus Assessment Group (BILAG) in 2004 as vasculitis or inflammation of small bowel, diagnosed on the basis of clinical features supported by suggestive radiological imaging and/or biopsy.[4] LE may be prevalent in about 0.2-9.7%,[5,6] of patients diagnosed with SLE but lupus enteritis presenting as the initial manifestation of the disease is quite rare.[7] On the other hand, diagnosing lupus enteritis without a prior background of SLE can be utterly challenging.

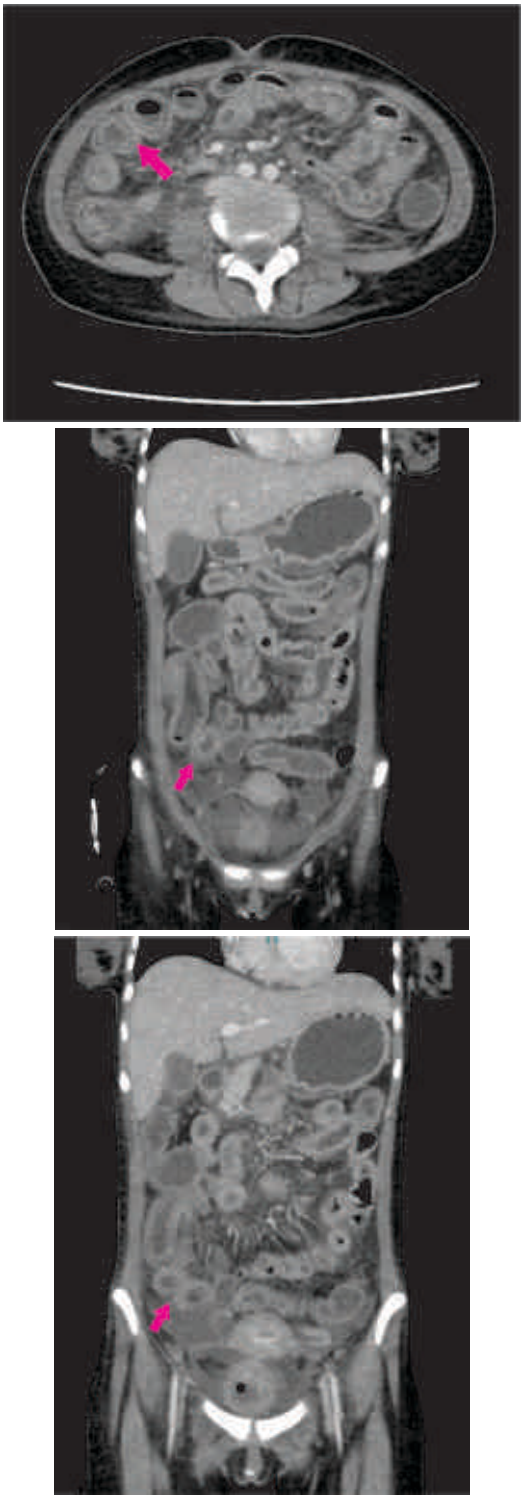
Case Presentation

A 21 years old Asian lady presented to the Emergency Department with the complaints of progressive generalized weakness associated with intermittent diarrhea and vomiting for past 6 weeks. She had been having 7-8 episodes of watery, non-bloody, large volume stools not associated with fever or abdominal pain. Vomiting was non-projectile, non-biliary and non-bloody, small volume, exaggerated in the last 4 days. She was initially seen on outpatient basis and prescribed antibiotics for these symptoms without any improvement in her clinical condition. She also reported a weight loss of about 7 kgs from her baseline weight. Her appetite had been well initially but she was unable to tolerate diet for past 4 days due to recurrent vomiting. She denied history of rash, joint pains, hair loss or oral ulcers. There was no recent travel history. She also denied having any allergy or addiction. There was no family history of tuberculosis or malignancy. Past medical history was significant for a second trimester abortion of unidentified cause about 2 months ago. She did not have any previous surgeries or blood transfusions. On examination, the patient was vitally stable. She was a lean built lady with vitiligo marks on her legs. She was alert and oriented to time, place and person. On chest auscultation, decreased air entry was observed in right lung base. Her abdomen was soft and non-tender without any visceromegaly. There was no shifting dullness and gut sounds were audible. Sensory motor examination was within normal limits. Laboratory investigations revealed anemia, severe

hypokalemia, hypomagnesemia, hypocalcemia and metabolic acidosis (Table.1) Based on history, clinical examination and these results, a differential diagnosis of celiac disease, inflammatory bowel disease, intestinal tuberculosis was made along with protein losing enteropathy associated with SLE or HIV. Further workup confirmed malabsorption syndrome, so she was planned for an upper and lower gastrointestinal endoscopy. Pre-procedure covid-19 PCR came out positive so the procedures were postponed. She underwent contrast enhanced CT scan abdomen which showed diffuse edema of stomach, small bowel and large bowel with positive target sign. Right sided pleural effusion and mild ascites were also noted. (Image.1) Meanwhile, her ANA, Anti-DsDNA, Lupus anticoagulant and anti-cardiolipin antibodies came out positive. (Table.2) The patient was diagnosed with Lupus enteritis with associated Anti-phospholipid syndrome. She fulfilled 8 of the 17 SLICC criteria including serositis, nephritis, hemolytic anemia, lymphopenia, presence of ANA, anti-DsDNA and anti-phospholipid antibodies as well as a positive direct COOMBS test. She was started on pulse therapy with IV methylprednisolone 1g per day for 3 days. Due to financial constraints, the patient was unable to continue further treatment in the hospital so she left against medical advice. She was discharged on oral prednisolone 30mg twice a day.

Table 1: *Baseline investigations of the patient.*

Test	Result	Reference range
Hemoglobin	6.0	11-14.5g/dl
Hematocrit	13.8	34.5-45.4%
Mean Corpuscular Volume	108.7	78.1-95.3fL
White blood cells	8.8	4.6-10.8 $\times 10^9$/L
Neutrophils	86.6	34.9-76.2%
Lymphocytes	7.8	17.5-45%
Platelets	339	154-433x10^9/L
Amylase	291	28-100IU/L
Creatinine	1.0	0.6-1.1mg/dL
Sodium	141	136-145mmol/L
Potassium	1.5	3.5-5.1mmol/L
Chloride	108	98-107mmol/L
Bicarbonate	16.9	20-31mmol/L
Calcium	7.6	8.6-10.2mg/dl
Magnesium	1.3	1.6-2.6mg/dl
Serum glucose	114	80-160mg/dl
Total bilirubin	3.0	0.1-1.2mg/dl
GGT	92	<38IU/L
ALT	47	<35IU/L
AST	126	<31IU/L
Alkaline phosphatase	173	45-129IU/L
Lipase	512	6-51U/L
PT	13.3	9.3-12.8seconds
APTT	20.4	22.9-34.5second



Images.1-3: *CT scan images showing diffuse bowel wall edema - target sign (See black arrow)*

Table 2: *Contains further blood workup of the patient.*

Test	Result	Reference Range
Stool analysis	+14 pus cells	
RBS folate	1300.4	280-791ng/ml
B12	>2000	>201 pg/ml Acceptable
LDH	715	120-246IU/L
ESR	47	0-20
Retic count	8.81	0.6-2.4%
COOMBs test	Positive (++++)	
TTG IgA	<0.5	Positive: >3.5 U/ml
TTG IgG	0.72	Positive: >3.5 U/ml
HIV	Non-reactive	
HbsAg	Non-reactive	
Anti-HCV	Non-reactive	
Albumin	2.5	3.5-5.2g/dl
Fecal Calprotectin	2.5	Negative <43.2ug/g
TSH	1.150	Adults: 0.4-4.2uIU/ml
ANA	Positive (Homogenous)	
Anti- DNA	1252	Positive >/= 25 IU/ml
Anti-SMQN	1.01	Positive > 5.0 U/ml
Anti-RO	1.27	Positive > 5.0 U/ml
Anti-LA	0.54	Positive > 12.5 U/ml
AntiSM	0.81	Positive > 5.0 U/ml
Anti Scl-70	1.43	Positive > 5.0 U/ml
Anti-cardiolipin IGM	23.41	Positive > 7.2 MPL-U/mL
Anti-cardiolipin IGG	15.40	Positive >14.4GPL-U/mL
LA screen	51.5	31-44 seconds
Spot Urinary Creatinine	69	mg/dl
Spot Urinary Protein	252	mg/dl

Limitations

Endoscopy should have been done as a part of workup for chronic diarrhea which was postponed due to a positive Covid-19 PCR result. Histopathological evidence was hence missing which could have added to the evidence pointing to our final diagnosis. Another limitation was lack of follow-up due to patient's social and personal issues, treatment response and need of additional immunosuppressive agents could not be evaluated.

Discussion

This case reports an uncommon presenting feature of a commonly occurring disease. It emphasizes that despite being rare, lupus enteritis is an important differential in patients presenting with gastrointestinal symptoms. It is crucial to identify and timely treat this condition as it may be associated with significant morbidity and mortality, hence the index of suspicion must be high.

SLE is a heterogenous disease in terms of its clinical presentation, evolution and prognosis but its manifestation with LE is exceptionally scarce. Lupus Enteritis was initially identified in 1980 and due to its heterogeneity and paucity of evidence; it has not been a part of the SLICC criteria for diagnosis of SLE.[9] Lupus Enteritis does not have characteristic clinical features and may present with diverse symptoms such as vomiting, abdominal pain, diarrhea, ascites or fever. The pathogenesis of disease is poorly understood but immune complex deposition along with complement activation seems to be the driving forces. The most commonly involved sites are jejunum and ileum followed by colon, duodenum and rectum respectively.[10,11] Endoscopy may not be helpful in such cases due to non-specific findings and the diagnostic yield of histopathology is barely 6%, hence CT abdomen with contrast is the gold standard for diagnosis of LE. Demonstration of bowel wall thickness greater than 3mm (Target sign), mesenteric vessels engorgement (Combs sign) and attenuation of mesenteric fat are the classical features of Lupus enteritis on imaging.[12] Management includes complete bowel rest and IV methylprednisolone as the initial therapy. If this is ineffective, immunosuppression with cyclophosphamide, mycophenolate or azathioprine should be considered.[13] Surgical intervention must be considered for resistant cases. Prognosis is usually excellent as the disease is fairly steroid responsive but relapse after initial improvement may be seen in upto 23% of cases.[9] Involvement of colon and bowel wall diameter > 9 mm have been illustrated as risk factors for recurrence.[14] In case of delay in treatment, LE may lead to bowel infarction, bleeding, obstruction and perforation.[7] A high fatality rate has been observed with the disease. Recently, Liu et al[15] have established a Lupus risk assessment model to predict the development of lupus enteritis in patients with SLE, its significance is still unclear, nevertheless, the model might turn out useful in early identification of the disease.

In a nutshell, this case highlights lupus enteritis as the initial manifestation of SLE and the key importance of CT scan in its diagnosis with a remarkable response to steroid therapy.

Conflict of Interest: *None*

Funding Source: *None*

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