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Systemic Lupus Erythematous Presenting As Intestinal Pseudo Obstruction: A Case Report.

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Abstract

Systemic lupus erythematous (SLE) is an autoimmune disease which involves multiple organs of our body and tissues. It has a varied clinical manifestation and at its onset, it may involve only one organ. We present a case of intestinal pseudo-obstruction as a rare presentation of SLE.

Case Presentation:

A 30year old female presented with pain abdomen for 3 days suggestive of dyspepsia with history of tiredness for 6 months. No history of rash, fever, joint pain, loose stools, vomiting, abdominal distension or jaundice. On examination, bowel sounds were sluggish, vitals were normal. X ray showed dilated bowel loops and ultrasound abdomen showed features of small bowel obstruction, CECT abdomen was done which was normal with Hepatomegaly. She was treated symptomatically. Routine Blood investigations showed anaemia with normal renal function, serum albumin was low with urine protein creatinine ratio of 3g, no evidence of any cast or haematuria and urine culture was negative. ANA profile showed, ANA positive with Anti-ds DNA, Anti-Ro, Anti-erythrocyte with low complement levels. Renal biopsy showed lupus nephritis stage V, she was started on anti-proteinuria drugs and HCQ along with high dose corticosteroids induction with MMF as maintenance and other supportive medications. She was followed up after 3 months and she was symptomatically better with her urine PCR being less then 1g/day.

Conclusion:

Intestinal Pseudo-obstruction, in this case, is a rare manifestation of systemic lupus erythematous.

Keywords: Systemic Lupus Erythematous, ANA Profile, Intestinal Pseudo-Obstruction

Introduction

Systemic lupus erythematous (SLE) is an autoimmune disease which involves multiple organs of our body and tissues. It has a varied clinical manifestation and at its onset, it may involve only one organ [1]. It is due to varied clinical manifestations many believe that this disease is a syndrome rather than a single disease. Symptoms of SLE can be musculoskeletal or musculocutaneous, renal, neurological, pulmonary and pleural disorders, blood disorders, heart and blood vessel disorders, and other organs [2,3].Intestinal pseudo-obstruction is a rare complication of systemic lupus erythematosus. Systemic lupus erythematosus-related intestinal pseudo-obstruction was initially reported in the 1970[1]. Asian populations are more likely to experience it, there is no mechanical obstruction, it typically manifests as signs and symptoms of large or small intestinal obstruction. As a result, the flow of abdominal contents is reduced and may be

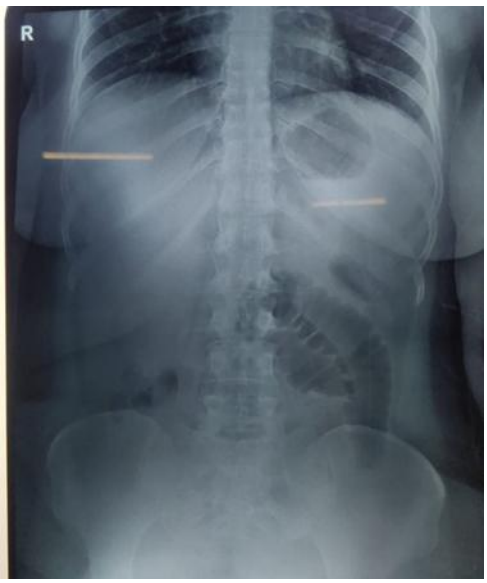
complicated by perforation, ischemia, and infarction [2, 3]. Only 1.96% of SLE patients experience IPO. Only about 57.6% of these 1.96% people had it as their first sign or symptom [2].

We hereby report a case of SLE with pseudo-intestinal obstruction which is rare.

Case Report

A 30year old female presented with pain abdomen for 3 days suggestive of dyspepsia with history of fatigue for 6 months. Her family history was not significant with no co-morbidities. No history of rash, fever, joint pain, loose stools, vomiting, abdominal distension or jaundice. No symptoms suggestive of cardiovascular, respiratory or renal involvement. On examination there was hyper-pigmentation of skin over the forehead and Physical examination revealed soft abdomen with sluggish bowel sounds and non-tender to palpation. Vitals were normal, laboratory results revealed anaemia with leukopenia, normal renal functions and normal liver enzymes with serum albumin of 2.5g/l. X ray showed dilated bowel loops (Figure 1) and ultrasound abdomen showed Distal ileal loops with prominently sluggish peristalsis to rule out intestinal obstruction, CECT abdomen was carried out, which was normal. It also showed grade 3 Hepatomegaly with fatty infiltration. She was treated conservatively.

Figure 1



She was evaluated for Hypoalbuminemia. Urinalysis showed protein of 3+. On further evaluation for proteinuria, Urine spot PCR showed 3g and 24hour urine protein showed 3.6g/day.

Further rheumatologic workup showed elevated antinuclear antibody (19 CTD-FEIA), double-stranded DNA antibody (>21 U/ml). Anti-Smith antibody was negative, erythrocyte sedimentation rate (ESR) was 70 mm/hr and C-reactive protein (CRP) was18.4 mg/L. It also showed low C3 complement 27 mg/dl, low C4 complement 4 mg/dl, as well as anti-SSA/Ro 60kD, anti-SSA/Ro 52kD (81 U/ml, 26 U/ml) respectively. Anti-Ribosomes was 85 U/ml, Anti-erythrocyte positive (Indirect Coombs test positive) and anti-Smith antibody were negative. Total WBC count was 3.67 K/μl with decreased absolute lymphocyte count of 0.72 K/μl.

Renal biopsy (Figure 2 & Figure 3) showed primary diagnosis of Lupus nephritis with pattern of injury being diffuse membranous glomerulonephritis, with mild tubulointerstitial chronicity and score/grade as ISN/RPS class V.

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Figure 2: Capillary loops are stiff, round and patent all throughout with thickening of basement membrane (40x PAS stain).

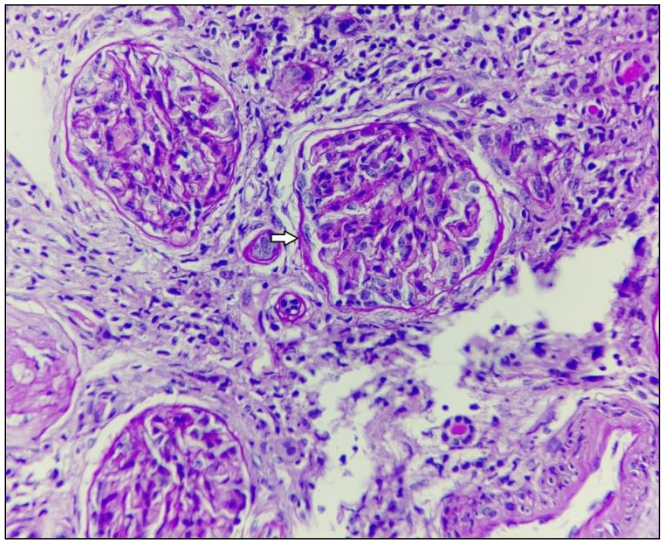
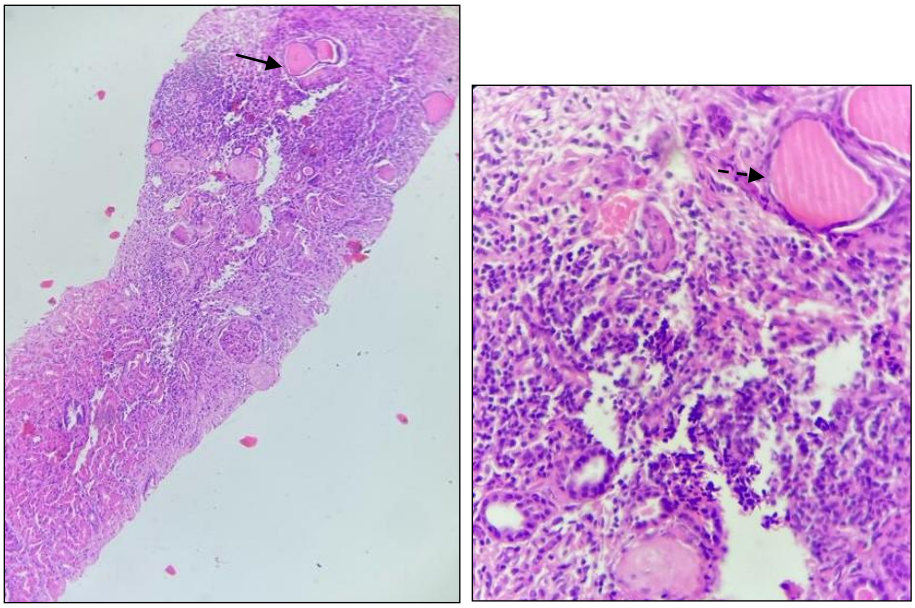


Figure 3: 10x H & E stain tubules showing hyaline cast in the lumen with mild degree of tubular atrophy / interstitial fibrosis.



The patient was treated with intravenous methylprednisolone 40 mg daily for 5 days and continued on maintenance steroid therapy which was slowly tapered and switched to oral methylprednisolone.

The patient was discharged home, on oral methylprednisolone and on subsequent follow up started on hydroxychloroquine 200 mg daily twice daily and mycophenolate mofetil 1 gm twice daily as steroid-sparing agent. SLE workup at the six-month clinic visit revealed urine protein of 0.7g/day, due to patient's refusal ANA profile was not repeated. All her bowel symptoms fully resolved and did not recur.

Discussion

Nausea, vomiting, and diarrhoea can be features of aggravation of SLE. The diffuse abdominal pain probably may be caused by autoimmune peritonitis and/or intestinal vasculitis. Gastrointestinal symptoms such as oesophageal hypomotility,

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malabsorption, mesenteric vasculitis, and pancreatitis are common in patients with SLE [4].

We present a rare case of IPO as one of the initial symptoms of active SLE. Zhang et al. discovered that, among 4331 SLE patients during the course of an 11-year longitudinal research, IPO was present in 1.96% of cases, with a 7.1% in-hospital death rate. With a misdiagnosis rate of 78%, IPO was the primary presenting cause in about 57.6% of these patients. Another two studies that took into account both newly diagnosed SLE and previously known cases indicated that IPO was the initial presenting symptom 50% and 41% of the time [2, 5].

Zhang et al. they found that, when compared, laboratory findings revealed that positive ANA, elevated CRP, hypoalbuminemia, hypocomplementemia, positive anti-SSA antibodies, and positive anti-SSB antibodies were more common in patients with IPO-related SLE [2]. Our patient's laboratory results were significant for a high titre antinuclear antibody (19- CTD Screen -FEIA), double-stranded DNA antibody (>21 U/mL), anti-SSA/Ro 60kD, anti-SSA/Ro 52kD (81 U/ml, 26 U/ml) respectively, anti-Ribosomes Po (85 U/ml), Anti-erythrocyte positive (Indirect Coombs test positive) and anti-Smith antibody.

Several theories have been offered in the literature regarding its pathogenesis. Due to the involvement of the digestive, hepatobiliary, and genitourinary systems, smooth muscle dysmotility is suspected secondary to vasculitis and immune complex deposition in the intestinal tract and bladder vessels [6, 7, 8].

A high clinical suspicion is required to diagnose IPO, which is a rare complication of SLE. It commonly involves the urinary tract with associated symptoms, and our patient did not have this involvement.

Conclusion:

Intestinal Pseudo-obstruction, in this case, is a rare manifestation of systemic lupus erythematous. Retrospectively we also diagnosed Lupus nephritis of stage V in our patient and treated her.

Female patients of reproductive age group who complain of fatigue, proteinuria and vague intestinal symptoms require further evaluation and look for treatable causes including SLE. Early diagnosis of SLE can prevent unnecessary surgical treatment.

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