Unusual manifestation of systemic lupus erythematosus

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ABSTRACT

The systemic lupus erythematosus (SLE) is a multisystem disease, and involvement of the gastrointestinal system is seen in about half of SLE patients. As a rare complication of SLE, acute pancreatitis presents as generalized flare-ups inpatients previously diagnosed with SLE. Lupus pancreatitis is more common in females and the third decade of life with an incidence ranging from 0.7% to 4%. Here, we report a rare case of acute pancreatitis as the initial presentation with later patient was diagnosed as a case of SLE, thus forming an unusual presentation of SLE.

Key words: *Acute pancreatitis, Anti-nuclear antibody, Autoimmune, Systemic lupus erythematosus*

The systemic lupus erythematosus (SLE) is a systemic autoimmune inflammatory disease with diverse clinical manifestations. Gastrointestinal tract (GIT) afflictions may be due to the disease itself or due to adverse reactions of medications or by opportunistic infections. GIT involvement in SLE has various forms such as mesenteric vasculitis (0.2–9.7%), protein-losing gastroenteropathy (1.9–3.2%), intestinal pseudo-obstruction which is rare, and lupus pancreatitis (0.7–4%) [1,2]. The annual incidence of acute pancreatitis in SLE is 0.4–1.1% [3-5]. Diagnosis of SLE pancreatitis is based on the clinical symptoms, laboratory tests, and temporal correlation. It is a rare presentation and very few cases of this entity are present in the literature.

Here, we are reporting the case of a 28-year-old female who presented with acute pancreatitis as the initial presentation and later diagnosed as SLE. By presenting this case, we aim to bring out the clinical characteristics of such patients. This will help clinicians to think of pancreatitis as an initial presentation of SLE and to keep it as one of the differential diagnoses of acute abdomen.

CASE REPORT

A 28-year-old female patient referred from Chamba Medical College with a history of fever with multiorgan dysfunction. The patient had a fever 15 days back which was high grade associated with chills, rigors, sweating, and intermittent without diurnal variations up to 101°F. The fever lasted for 6 days and the patient was afebrile for 9 days before she presented to our hospital. The patient also had myalgias along with fever. There was a history of yellowish discoloration of eyes for the past 15 days along with pain in the abdomen which was radiating to the back.

On examination, blood pressure was 136/78 mmHg, pulse rate was 90/min regular, good volume without any special character, and respiratory rate was 16/min. Pallor was absent and there was no cyanosis. Icterus was present and there was pitting edema. The jugular venous pressure was not raised. There was no palpable lymphadenopathy. Cardiovascular, central nervous system, and respiratory system examination were within normal limits. Abdominal examination showed diffuse tenderness with shifting dullness and there was no lump palpable. No organomegaly was present.

On investigation, the patient's hemoglobin was 10.4 g/dl, total leukocyte count (TLC) was 4860/mm^3 (neutrophils – 89% and lymphocytes – 9%). TLC done on the 3rd day was 2830/mm^3. The platelet counts were 103,000/mm^3. Her mean cell volume was 84.42 fl and the mean hemoglobin concentration was 29.46 pg. Peripheral smear showed normocytic normochromic anemia with thrombocytopenia. Bilirubin (total) was 6.12 mg/dl, direct was 3.5 mg/dl. Serum aspartate transaminase was 332 U/L, alkaline phosphatase was 92, and creatinine was 3.43 mg/dl. Her total proteins were 6.1 g/dl and albumin was 2.2 g/dl. Serum electrolytes were in the normal range. Her serum amylase was 536 U/L and serum lipase was 492 U/L.

Fever workup showed IgM scrub to be negative, IgM leptospira negative, and dengue NS1 negative. Enzyme-linked immunosorbent assay for human immunodeficiency virus and hepatitis B and C viruses were all negative. The ascitic tap was done and the analysis showed that the appearance was straw and clear, total protein was 2 g/dl, and albumin was 1 g/dl, while serum protein was 5.4 g/dl, and serum albumin was 2 g/dl, serum albumin ascitic gradient was 1 (low), glucose was 101 mg/dl, TLC was 59 (N 10% and L 90%), red blood cell was 170, and ascitic fluid

amylase was 303 mg/dl (high). Urine albumin creatinine ratio was 214 mg/g of creatinine. There were no urinary casts. Serum procalcitonin was 0.343. The direct Coombs test was negative. C-reactive protein was 6 mg/dl. Serum leukocyte dehydrogenase was 1025 U/L and Vitamin B12 was >2000 ng/ml. Blood cultures were sterile and upper gastrointestinal endoscopy was normal. Thyroid function tests showed T3 – 38.4 ng/ml, T4 – 2.86 mcg/dl, and TSH 13.151 mcIU/ml.

The contrast-enhanced computed tomography (CECT) abdomen and pelvis showed gross ascites, bilateral pleural effusion (with passive atelectasis of underlying lung parenchyma), and anasarca with roundworms in the jejunal loops with small-sized uterus (Figs. 1 and 2).

The patient was initially managed on the lines of pyrexia with multiorgan dysfunction syndrome keeping the possibilities of scrub typhus and leptospirosis. Serum amylase and lipase were raised. Ascitic fluid amylase was also increased. Diagnosis of acute severe pancreatitis (BISAP 2/5) was kept. In view of pancytopenia, autoimmune etiology was kept for acute pancreatitis. Anti-nuclear antibody was sent which came out to be positive (Hep 2 – 1:1250 dilution) and workup for SLE was done. During the hospital stay, the patient had the first episode of generalized seizure on the 15th day of admission.

The Systemic Lupus International Collaborating Clinics (SLICCS) criteria were positive in this patient. There were six clinical and four immunological criteria positive in our case (Table 1). Hence, a diagnosis of acute pancreatitis with SLE was kept in view of SLICCS criteria. A final diagnosis of SLE with pancreatitis was made and the cause for pancreatitis was kept as SLE because of temporal correlation as well as no other cause of pancreatitis was found. The patient was started on steroids 1 mg/kg body weight for 6 weeks followed by tapering of steroids. Over the next 4 weeks, the pain of the patient improved as well as ascites and fat stranding. The patient is under regular follow-up.

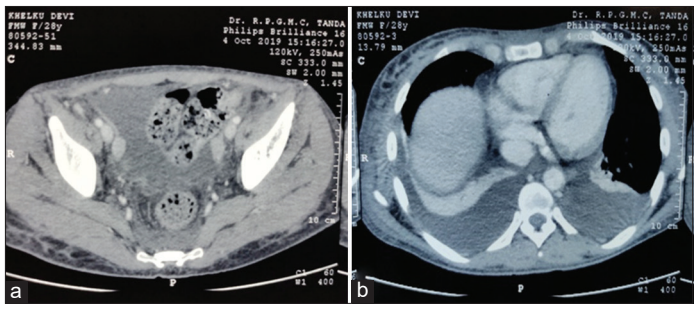

Figure 1: Contrast-enhanced computed tomography abdomen showing (a) gross ascites and (b) bilateral effusion

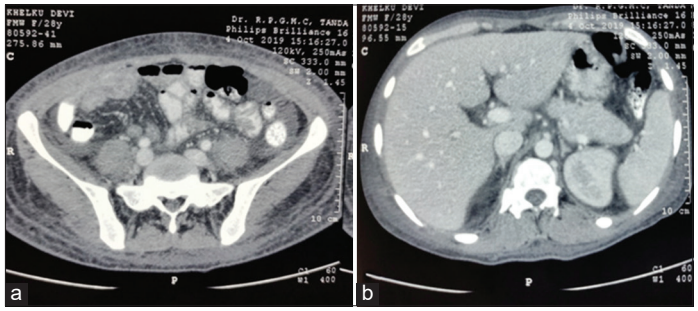

Figure 2: Contrast-enhanced computed tomography abdomen showing (a) mesenteric fat stranding along with round worm in the jejunal loops; (b) bulky body of the pancreas

Table 1: SLICCS score of our patient

Positive clinical manifestations	Positive immunological manifestations
History of oral ulcers	ANA – 1:1250 end point titer (homogenous pattern)
Non-scarring alopecia	Anti-dsDNA – positive (1:10 end point titer)
Seizure	Low C3 – 50 mg/dl
Anemia	Low C4 – <7.3 mg/dl
Leukopenia	
Thrombocytopenia	

ANA: Anti-nuclear antibody, SLICCS: Systemic Lupus International Collaborating Clinics

DISCUSSION

Common precipitants of acute pancreatitis include hepatobiliary tract disease-related mechanical obstruction and metabolic insults including alcohol, drugs, hypercalcemia, and hypertriglyceridemia. Around 20% of cases of acute pancreatitis are deemed idiopathic. Pathogenic features of SLE pancreatitis may include vasculitis, microthrombus formation, anti-pancreatic antibodies, and inflammation due to T-cell infiltration, and complement activation [6-9].

The diagnosis of acute pancreatitis is based on clinical symptoms and pancreatic enzyme elevation and may be supported by characteristic imaging findings. However, SLE patients can have subclinical presentations of pancreatitis, with an elevation of pancreatic enzymes in the absence of clinical symptoms [6,8]. Another distinguishing feature of lupus pancreatitis is that it can present in association with cytopenia. Anemia, leukopenia, and thrombocytopenia occur in 81%, 59%, and 48% of cases, respectively, while leukocytosis is infrequent (15%) [3]. SLE pancreatitis can occur as a generalized flare or during disease quiescence.

Pathophysiology of SLE-related pancreatitis is thought to be multifactorial and the exact mechanism is not yet known. There are certain risk factors associated with SLE-related pancreatitis – hypertriglyceridemia, recent viral infection, psychosis, and drug toxicity are the important ones [10]. Complement activation and autoimmune reactions, viral infections, vasculitis, intimal thickening, proliferation, and immune complex deposition may play a role in the underlying mechanism of the disease [7]. Autoantibody production along with abnormal cellular immune response may lead to inflammation of parenchyma [1]. The increase in the number of cases of pancreatitis as an initial presentation of SLE supported that SLE is the underlying etiologic factor [7]. Hence, in this case, the patient had typical pain abdomen, biochemistry, and CECT abdomen findings suggestive of acute pancreatitis along with positive SLICC

criteria. Interestingly in our patient, the CECT abdomen also showed roundworms in the jejunal loops.

Ascaris is one of the most common intestinal parasites in the world. About 25% of the world's population is infected with this helminth [11]. The life cycle of *Ascaris lumbricoides* starts after the ingestion of the egg. After hatching, larvae migrate to the lungs through the intestinal wall through the portal and systemic circulation, penetrate the alveolar walls, ascend the bronchial tree to the throat, and are swallowed into the small intestine again, where they mature into adult worms [12]. The disease may stay asymptomatic with ascariasis in the lumen of the small intestine. Symptoms can occur when helminths invade the biliary or pancreatic ducts. Presentations of forms are biliary colic (56%), acute cholangitis (25%), acute cholecystitis (13%), and acute pancreatitis (6%), and, rarely, hepatic abscess or hemobilia [13]. Hence, ascariasis can be a remote possibility of acute pancreatitis in this case. The patient was managed conservatively for acute pancreatitis and was started on oral prednisolone for SLE and albendazole was given for ascariasis which resulted in the clinical improvement in our case.

CONCLUSION

SLE presenting as pancreatitis is a severe form of SLE which is rarely seen in clinical practice. Early diagnosis and timely initiation of steroid pulse therapy may improve the outcome in SLE patients presenting with pancreatitis. Steroid therapy in acute pancreatitis of other etiology is hazardous. It is also difficult to treat SLE pancreatitis with steroids because of the known toxic effects of steroids, but they can make remarkable improvements in the prognosis of such patients due to their immunosuppressive effects. If it is SLE-related pancreatitis, after ruling out other causes of pancreatitis, the steroid is advised to be started early for better outcome of the patient.

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