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Case Report

Lupus Enteritis: An Uncommon Manifestation of Systemic Lupus Erythematosus as an Initial Presentation

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Abstract

Systemic lupus erythematosus (SLE) is an autoimmune disorder generally affects young to middle-aged women, commonly presenting as a triad of fever, rash, and joint pain but can affect multiple organs and can present in a complex fashion, varying based on the degree and severity of organ involvement. The differential for abdominal pain and diarrhea in SLE is vast and can include VIPomas, serositis, pancreatitis, intestinal vasculitis, and protein – losing enteropathy, gluten – enteropathy, intestinal pseudo-obstruction, and infection. The pathology of lupus enterits thought to be immune-complex deposition and complement activation, with subsequent mucosal edema. We present a case of a woman with no history of SLE, but with a prolonged course of abdominal pain, diarrhoea and vomiting and eventual diagnoses of lupus enteritis.

Keywords: *Lupus erythematosus, systemic lupus erythematosus*

Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disorder generally affects young to middle-aged women, commonly presenting as a triad of fever, rash, and joint pain but can affect multiple organs and can present in a complex fashion, varying based on the degree and severity of organ involvement. The differential for abdominal pain and diarrhea in SLE is vast and can include VIPomas, serositis, pancreatitis, intestinal vasculitis, and protein-losing enteropathy, gluten enteropathy, intestinal pseudo-obstruction, and infection.[1] The pathology of lupus enteritis thought to be immune complex deposition and complement activation, with subsequent mucosal edema.[1] We present a case of a woman with no history of SLE but with a prolonged course of abdominal pain, diarrhea and vomiting, and eventual diagnoses of lupus enteritis.

Case Report

A 20-year-old unmarried female with no significant medical history presented with complaints of diffuse, constant, dull aching abdominal pain for 20 days, associated with loose stools and vomiting. Loose stool had a frequency of 8–10 episodes per day, watery in consistency and associated with vomiting 2–3 bouts per day nonbilious and contained ingested food. She was treated with metronidazole and ciprofloxacin outside for presumed infectious colitis but did not show much improvement. She presented to our hospital with persistent similar complaints and was admitted for further evaluation. On physical examination, vitals were within normal limits. The abdomen revealed mild diffuse tenderness on deep palpation and shifting dullness on percussion. Rest of the systemic examination was within normal limits. Initial possibility of intestinal and mesenteric tuberculosis with ascites was kept, and diagnostic paracentesis was performed. Twenty milliliters of straw-colored ascetic fluid aspirated and analysis revealed high protein (3.5 g/dl), low serum-ascites albumin gradient (0.8), and neutrophil predominant ascites (total cells 350 with 65% neutrophil and 35% lymphocytes), with adenosine deaminase (ADA) of 18 U/L. As ascetic fluid analysis was not suggestive of tuberculosis (low ADA and neutrophil predominant ascites), contrast-enhanced computed tomography (CT)

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of abdomen was performed which [Figure 1a and b] revealed marked bowel wall edema involving small and large bowel ("target sign"), dilatation of the intestinal segments, engorgement of the mesenteric vessels ("comb sign"), increased attenuation of the mesenteric fat, and ascites. In view of bowel wall thickening on imaging, video-colonoscopy [Figure 2a and b] was performed which revealed edematous mucosa with mild hyperemia in proximal ascending colon, cecum, ileocaecal valve, and terminal ileum without any ulceration, friability, granularity, and nodularity. Intestinal biopsy was taken which showed normal histology. *Mycobacterium tuberculosis* not detected in polymerase chain reaction for the detection of *M. tuberculosis* in ileal biopsy specimen.

In view of diffuse bowel involvement, characteristic imaging findings, and absence of changes suggestive of intestinal tuberculosis and inflammatory bowel disease in ileocolonoscopy, possibility of lupus enteritis was kept, and workup for autoimmune disease was started.

Laboratory tests revealed a positive antinuclear antibody (ANA) titer of 1:1280, low complement levels, negative serology for anti-dsDNA, positive direct and indirect Coombs test, and leukopenia. Urine examination revealed dipstick 2+ proteinuria, and 24 h urine examination revealed 6.6 g/24 h proteinuria. Hepatitis B, hepatitis C, and HIV serology were negative.

A kidney biopsy was performed which on direct immunofluorescence and periodic acid-Schiff staining showed mesangial hypercellularity and mesangial deposits with the expansion of the mesangial matrix suggestive of International Society of Nephrology Class II lupus nephritis [Figure 3]. Due to nephrotic-range proteinuria, possibility of lupus podocytopathy was kept.

Presence of cardinal signs on CT scan along with positive ANA and evidence of lupus nephritis on renal biopsy led to the correct diagnoses of lupus enteritis. The patient was treated with high-dose (1000 mg) intravenous (IV) "pulse" methylprednisolone daily for 3 days with Euro-Lupus IV cyclophosphamide regimen, which resulted in significant clinical improvement. Patient showed rapid improvement with resolution of abdominal pain, vomiting, and diarrhea. Need for cyclophosphamide reassessed in consultation with nephrologist, and in view of rapid improvement with pulse steroid and single infusion of cyclophosphamide, it was decided to discontinue cyclophosphamide and patient discharged on oral steroid, angiotensin-converting enzyme, inhibitors, and calcium supplements. On follow–up, there is resolution of ascites and patient started on tapering doses of steroids.

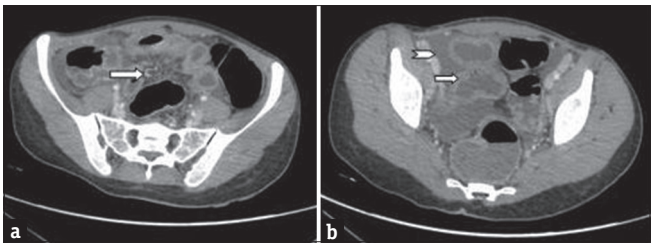

Figure 1: (a) Mesenteric hypervascularity (arrow), (b) Target sign (arrow) and ascites (arrowhead)

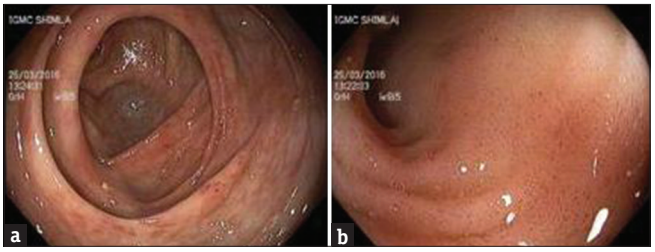

Figure 2: (a) Edematous mucosa with mild hyperemia in caecum and proximal ascending colon and (b) edematous mucosa with mild hyperemia in terminal ileum

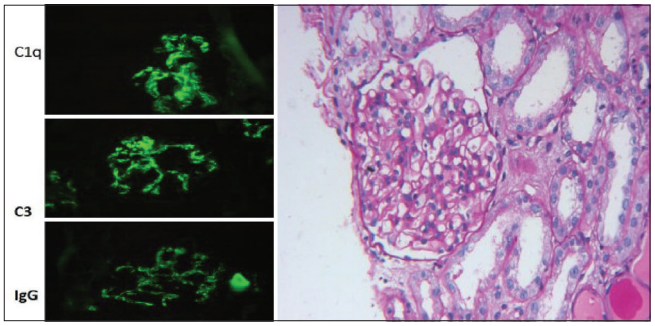

Figure 3: Direct immunofluorescence images show 2+/3+ mesangial granular and confluent staining for their respective immunoreactants (C1q, C3, and IgG). Periodic acid-Schiff staining show mesangial expansion with few areas of mesangial hypercellularity and stiffness of capillary basement membrane

Discussion

Lupus enteritis, also termed as mesenteric arteritis, lupus mesenteric vasculitis, lupus arteritis, and lupus vasculitis, was first described by Hoffman and Katz[2] in 1980. In the BILAG 2004, lupus enteritis is defined as either vasculitis or inflammation of the small bowel, with supportive image and/or biopsy findings.[3] Mean age at diagnosis of lupus enteritis reported in literature is 32.5 years, with the youngest patient being 13 and the oldest being 72-years old and male–female ratio of 1:14.[4] In a recent review of literature by Janssens *et al*.,[5] on 150 patients of lupus enteritis revealed abdominal pain (97%) as the most common symptom, followed by ascites (78%, clinical or radiological), nausea (49%), vomiting (42%), diarrhea (32%), and fever (15/74; 20%). Lupus enteritis is seldom confirmed on histology. Only 6% had clear histological confirmation

of vasculitis and[5] 7% had complications in the form of intestinal necrosis or perforation, yielding a mortality rate of 2.7%. Typically, the abdominal pain described in lupus enteritis is diffuse in pattern and in some cases accompanied by rebound tenderness.[6] Other than a drop in white blood cell count and complement titers, which may correlate with the occurrence of lupus enteritis, none of the laboratory indices have been found to be useful to establish the diagnosis.[4] The patient discussed had leukopenia and low complement levels although proof of vasculitis was not found in our case.

Lupus enteritis leads to three cardinal imaging signs on abdominal CT: (1) bowel wall thickening (target sign), ascites, and dilatation of intestinal segments, (2) engorgement of mesenteric vessels ("comb sign"), and (3) increased attenuation of mesenteric fat.[7] Target sign is "thickened bowel wall with enhancing outer (muscularis propria/serosa) and inner (mucosa) layers and a hypoenhancing middle layer due to submucosal edema." Our patient's CT findings were classic and were instrumental in making the correct diagnosis.

Bowel ischemia due to lupus enteritis has been shown to be less ominous than previously expected, and the abnormal CT findings were reversible with early diagnosis and prompt treatment.[8] Lack of specificity of these signs is a limitation of CT because they can also be seen in patients with pancreatitis, mechanical bowel obstruction, peritonitis, or inflammatory bowel disease, all of which may mimic intestinal ischemia[4,6]

There are no randomized control trials on treatment of this entity. Lupus enteritis has been found in several reports to be generally reversible and steroid responsive, thus considered as the first-line therapy.[4,6] There is preference for IV steroid in case of severe lupus flare because of potentially reduced drug absorption due to enteritis.[9] Treatment can be switched to oral corticosteroids as soon as adequate clinical improvement occurs. Hydroxychloroquine, mycophenolate mofetil, azathioprine, and low-dose corticosteroids could be considered for long-term maintenance treatment although it is unknown whether they may prevent recurrences. Cyclophosphamide or mycophenolate may be added in case of resistance to corticosteroids or when warranted by other organ involvement.[6] In our case due to associated renal involvement, patient started on pulse IV methylprednisolone along with Euro-Lupus low-dose IV cyclophosphamide regimen. Patient improved with resolution of abdominal pain, vomiting, and diarrhea and patient advised for close follow-up.

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Conflicts of interest

There are no conflicts of interest.

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