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Case Report
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Lupus Enteritis in Systemic Sclerosis-Systemic Lupus Erythematosus Overlap Syndrome

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Abstract

Lupus enteritis has not been well described in patients with overlap syndrome. It is rare both as an initial presentation of overlap syndrome and as a flare in known disease. We describe a case of lupus enteritis in a female patient with known systemic sclerosis-systemic lupus erythematosus overlap syndrome presenting with severe abdominal pain and non-specific GI symptoms. Diagnosis was made clinically and with CT imaging. A high index of suspicion should be maintained for lupus enteritis in patients with overlap syndrome due to high mortality associated with lupus enteritis. The main stay of treatment is immunosuppression and steroid therapy.

Keywords: Lupus enteritis; Immunosuppression; Steroid therapy

Introduction

Lupus enteritis has not been well described in patients with overlap syndrome. It is rare both as an initial presentation of overlap syndrome and as a flare in known disease. Overlap syndromes are rheumatologic disorders that satisfy criteria for 2 or more rheumatologic diseases, most commonly involving combinations of systemic sclerosis, systemic lupus enteritis, rheumatoid arthritis, dermatomyositis, and polymyositis [1]. Patients with Systemic Lupus Erythematosus (SLE) often have gastrointestinal symptoms and can develop the rare complication of lupus enteritis [2]. The incidence ranges between 0.2% to 5.8% [1]. We report a case of a 21 year old female with overlap syndrome including SLE, RF positive polyarticular arthritis, interstitial lung disease, and severe Raynaud's disease who presents with acute severe abdominal pain and found to have lupus enteritis.

Case Presentation

We present a 21 year old female with Overlap syndrome including SLE, RF positive polyarticular arthritis, interstitial lung disease, and severe Raynaud's disease. Her rheumatologic markers included a high titer ANA. She had positive Smith, RNP, and SSA antibodies as well as low positive dsDNA antibodies, elevated ribosomal P antibody, hypocomplementemia (C3 and C4), and indeterminate antiphospholipid antibodies. She also met incomplete criteria for Systemic Sclerosis, concerning for evolution of her disease.

She presented acutely with generalized abdominal pain, non-bloody diarrhea and intractable vomiting. Differential diagnosis included pancreatitis, toxic ingestion, medication side effect, viral gastroenteritis, cannabis hyperemesis syndrome, peritonitis, ascites, irritable bowel syndrome, autoimmune hepatitis, gastrointestinal dysmotility. Lupus can also rarely cause a vasculitis of the intestines called Lupus enteritis. She was found to have lupus enteritis diagnosed using CT of the abdomen and pelvis, performed due to her severe abdominal pain. Lupus enteritis presents in young women with SLE between puberty and menopause. Making an early diagnosis of Lupus enteritis is important as it is associated with a mortality rate that approaches 11% when the patient presents with acute abdominal pain [1]. Etiology of Lupus enteritis is attributed to a Lupus related mesenteric vasculitis resulting in intestinal pseudo-obstruction and protein losing enteropathy. Oral ulcers typically are simultaneously present [1]. This patient did not have oral ulcers; instead she had midline suprapubic abdominal tenderness but otherwise with soft and non-distended abdomen. Her home medications included mycophenolate, hydroxychloroquine, aspirin and nifedipine but had been non-adherent to medications for the month preceding presentation. She had not had an autoimmune disease flare for three years prior to this acute presentation.

Discussion

In a review of lupus enteritis radiologic findings, the most frequent abnormalities included bowel

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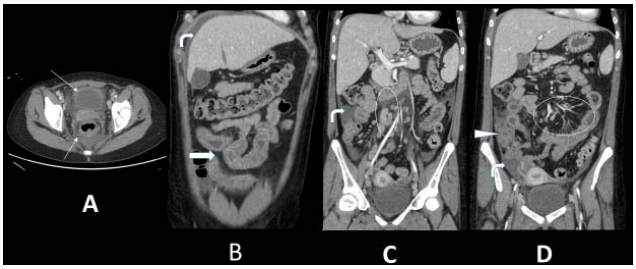

Figure 1: Axial (A) and Coronal (B-D) CT images of the abdomen and pelvis performed after contrast administration showed thickening of the rectal wall with adjacent inflammatory changes and questionable thickening of the bladder wall (thin white arrows-A); Small bowel enteritis with "target sign" (thick white arrow-B); Ascites (curved white arrows-B,C,D); probable duodenitis with adjacent fluid and retroperitoneal adenopathy (tall white oval C); mesenteric vasculature was engorged near the enteritis in several locations (flat white oval-D). There was some wall thickening of the cecum (white arrow head-D).

wall edema, bowel wall enhancement (double halo/target sign), and dilation of bowel lumen. Ascites was present in up to 78%. Mesenteric abnormalities including mesenteric vessel engorgement, increased number of visible vessels (comb sign) and increased mesenteric fat attenuation was present in 71%3. CT scan remains the gold standard for lupus enteritis diagnosis (Figure 1).

The most common laboratory findings of lupus enteritis include hematologic derangements (leukopenia, lymphopenia, and anemia), positive ANA (92%), anti-dsDNA (74%), low complement (88%), anti-RNP (28%), anti-SSA (26%), and anti-Sm (24%) [2]. CRP elevation is not characteristic of the disease [3].

In the patient's hospital course, her laboratory markers supported a rheumatologic flare. She had a low CRP but markedly elevated ESR. She had a normal CBC, uric acid, and mildly elevated LDH that was reassuring against oncologic process. Lipase, urine culture, stool calprotectin, liver enzymes, stool hemoccult and stool pathogen panel were negative, reassuring against infectious and other acute abdominal etiology. She was treated with pulse steroids and monitored closely. She was also restarted on her home medications. Her course was complicated by development of pulmonary edema that improved given furosemide, however her abdominal pain resolved and was discharged home on oral steroids.

Conclusion

Lupus enteritis is known to be a rare presentation of SLE flare [3]. Our patient with a known SLE component of her overlap syndrome demonstrated severe acute GI symptoms along with characteristic radiographic findings of Lupus enteritis that subsequently improved with standard rheumatologic flare treatment. This patient's medication non-adherence may have contributed to her flare. It is not documented in the literature whether overlap syndromes with SLE components have increased risk of lupus enteritis compared to patient's with SLE alone. Just as in isolated SLE patients who present with non-specific abdominal pain, there should be a high index of suspicion for lupus enteritis in patients with overlap syndrome and non-specific GI symptoms. Immunosuppression and steroids remains the main stay of treatment [2].

References

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