Systemic lupus erythematosus with intestinal pseudo-obstruction: a case report and review of the literature

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Abstract

Intestinal pseudo-obstruction is a rare complication of Systemic Lupus Erythematosus (SLE). Due to diagnostic challenges, it can go largely unrecognized. It is characterized by features of mechanical obstruction of the small or large bowel in the absence of an anatomic lesion obstructing the flow of intestinal contents. There are a paucity of case reports in literature worldwide. We report two cases of intestinal pseudo-obstruction in which one patient survived. The two cases illustrate the need for a high index of suspicion for diagnosis and early intervention with systemic high dose corticosteroids and IVIG. This will avert surgical intervention and reduce the mortality rate from this manifestation of lupus.

Key words: Systemic lupus erythematosus, Intestinal pseudo-obstruction

Introduction

Systemic Lupus Erythematosus (SLE) is a chronic, inflammatory, autoimmune disease characterized by multisystemic involvement with a wide variety of clinical presentations. Gastrointestinal complications are commonly overlooked in SLE[1]. They constitute about 40% of lupus manifestations. Approximately 2-30% of the complaints are due to the disease while the remainder due to adverse reactions to the medicines of lupus. Other manifestations are due to vasculitis, infections or other intercurrent processes (e.g. uremia)[1]. The manifestations range from vomiting, diarrhoea to protein-losing enteropathy and mesenteric vasculitis. Intestinal pseudo-obstruction is rare and seen in up to 23% of gastrointestinal manifestations[2]. We report two cases of intestinal pseudo-obstruction. We compare our two cases with results of a literature review looking at presentation, treatment, and outcomes.

Case report 1

A 59-year-old lady with lupus nephritis referred with complaints of vomiting, abdominal pain, and constipation but was passing flatus. She was reported to have had episodes of haematemesis. She was known lupus nephritis on mycophenolate, hydroxychloroquine, and prednisone. Upper GIT endoscopy revealed Mallory Weiss tear. On admission, she was noted to have a deranged renal function in keeping with acute kidney injury. She was commenced on prokinetics and hydration with intravenous fluids. However, the complaints of abdominal pain vomiting and constipation had progressed to no stool for three days. CT scan abdomen revealed dilated loops of small and large bowels, suggestive of intestinal obstruction, ascites, and basal pleural effusion. She also had a low complement, deranged renal function, and low haemoglobin. The diagnosis of SLE flare with intestinal pseudo-obstruction was made based on clinical findings CT scan findings that revealed dilated bowels without evidence of an abrupt transition point or mechanically obstructing lesion. A decision to start 5 days pulse with methylprednisone was made. During the treatment, she developed steroid-induced psychosis which later resolved following review by a psychiatrist. She improved the inflammatory markers came down and she opened bowels. Two days later she developed abdominal distention, was now vomiting and not passing stool or flatus. Her complement levels decreased and her DsDNA levels went up. A decision to pulse again with high dose steroids was made as efforts to obtain IVIG was underway. She, however, developed sepsis during the treatment and succumbed.

Case report 2

A 27-year-old lady with lupus nephritis was admitted with diarrhoea and vomiting. The test confirmed the diagnosis of cholera for which antibiotics and intravenous fluid was commenced. The initial kidney tests were deranged with proteinuria of 1+

and haematuria 1+. The patient improved with vomiting reducing and had started forming solid stool though of a small amount for the next 4 days. On day 7 the vomiting recurred and despite the treatment, it was noted that vomiting persisted and her creatinine was rising. At this point, she reported that she hadn't passed stool or flatus for two days. Abdominal X-ray and CT scan showed dilated loops of small bowels, suggestive of intestinal obstruction, mild ascites, and small pericardial effusion. She was noted to have low complements and rising titers of DsDNA. The diagnosis of intestinal pseudo-obstruction was made based on clinical findings and the fact that the CT abdomen failed to show abrupt transition point or mechanically obstructing lesion despite having dilated large intestines. At this point decision to pulse with 1gm of methylprednisone for was made. The gastrointestinal symptoms gradually improved and the SLE went into remission.

Discussion

Intestinal Pseudo-Obstruction (IPO) is a rare and commonly missed or underdiagnosed manifestation of lupus. In a review by Wang *et al*[3] on 40 cases of IPO, they noted a female to male ratio 12.3 with a mean age of 32.5 (10–57) years. Our two patients were female aged 60 and 27 years. The most common symptoms include abdominal pain (80%), vomiting (78%), diarrhoea and/or constipation (70%) and abdominal distension (63%)[3]. Misdiagnosis is often common especially if it's the presenting complaint of lupus. This can lead to unnecessary surgery because of being misdiagnosed[3]. The mortality from SLE related IPO can range from 25% to as high as 67%[4]. Case one had a delay in diagnosis and may be this may have contributed to the mortality. The diagnosis was made only after the vomiting and constipation persisted. Our threshold for diagnosis in the second patient was higher especially after our encounter with case 1.

The pathogenesis of SLE-related intestinal pseudo-obstruction is still not well understood. The possible mechanisms include immune complex deposition of anti-ds-DNA antibodies and C1q among others in smooth muscle cells, or vasculitis leading to damage, chronic ischaemia and ultimately hypomotility of the intestines[4,5]. Diagnosis is based on symptoms and tests. There will be evidence of systemic disease activity like decreased serum levels of complement, positive titers of anti-dsDNA, Sjögren's syndrome type A antibodies (anti-Ro) and anti-ribonucleoprotein (RNP) antibody and negative Sjögren's syndrome type B antibodies (anti-La). Both patients had decreased serum levels of complement and positive titers of anti-dsDNA. Our second patient had positive titers for Sjögren's syndrome type A antibodies (anti-Ro)[6,7]. Radiological investigations (abdominal X-ray/CT scan) can reveal a grossly dilated bowel; abnormal bowel gas patterns; thickening of the intestinal walls, signs of peritonitis, pyelocaliectasis, and ureterohydronephrosis[8]. Both our patients developed intestinal pseudo-obstruction in the background of lupus nephritis. The reasons as to what would predispose its development probably electrolyte derangements, but further research is still needed.

Once the diagnosis is established, the high dose corticosteroids are recommended as first-line treatment[3-8]. Role of surgery is very minimal. In a review by Wang *et al*[3], they reported that all patients who had surgery as first-line treatment relapsed. It was also noted these patients responded to a trial of corticosteroids following the relapse[3]. Case 2 responded to the steroid pulse. Case 1 did not respond, she also developed sepsis post the steroid and thus limiting our second line options. The second line includes IVIgG, cyclophosphamide, and tacrolimus[3,9]. IVIgG is not easily available in our set up due to its cost and the presence of sepsis ruled out cyclophosphamide in case 1. Other measures, such as nutritional support and prokinetic agents used in other causes of intestinal pseudo-obstruction, can be used in management.

Conclusion

Clinicians should have a high index of suspicion of IPO secondary to lupus. This will ensure quick accurate diagnosis and initiation of the appropriate treatment. There could be a link between nephritis and development of intestinal pseudo-obstruction in lupus. Outcomes of patients with IPO secondary to lupus who have undergone surgery and invasive procedures have been disappointing. Evidence points towards quick initiation of immunosuppressants such as steroids as the first line with IVIG as second-line options. This will avert unnecessary surgical intervention and reduce the mortality rate from this manifestation of lupus.

Conflicts of Interest

No potential conflict of interest relevant to this article was reported.

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