Research Article

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A report of chronic intestinal pseudo-obstruction related to systemic lupus erythematosus

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Abstract: Chronic intestinal pseudo-obstruction (CIPO) is a functional gastrointestinal disorder with symptoms of ileus. CIPO can either be idiopathic or secondary to other diseases such as systemic lupus erythematosus (SLE). SLE is involved in many parts of the gastrointestinal system with variable clinical presentations. Reports about complicated CIPO as a complication of SLE is infrequent. A 49-year-old female suffering from clinical symptoms of ileus has been hospitalized 3 times over 1 year. Her examination results showed no observation of mechanical obstruction. In August 2017, she came to the nephrology department due to edema in both lower limbs along with symptoms of ileus. After thorough examination, she was diagnosed with secondary CIPO related to SLE. Results of renal biopsy confirmed to be lupus nephritis (Class III-(A) + V). The symptoms of ileus are gradually improved after treatment of full-dose intravenous corticosteroid for 5 days.

Keywords: Systemic lupus erythematosus; Intestinal-pseudo obstruction; Renal Biopsy; Intestinal Biopsy

2 Case report

A 49-year-old female was admitted to our hospital in August 2017 with abdominal pain and distension, accompanied by edema in lower limbs, nausea, vomiting, no venting and defecation for 4 days. She was formerly hospitalized twice in July 2016 and May 2017 for similar milder symptoms. She was diagnosed with functional ileus because enteroscopy showed no evidence of mechanical obstruction. She was given supportive treatment including gastrointestinal decompression and coloclyster. The supportive treatment relieved her symptoms gradually, such as vomiting, abdominal pain and distension. For the third time she got same treatments at a local hospital, but the symptoms continued to worsen. So she came to our department for further treatment. Physical examinations showed multiple decayed teeth, abdominal distension, middle abdominal tenderness, absence of bowel sound, no facial rush. Laboratory examination revealed proteinuria, hypoalbuminemia, increased neutrophil proportion, antinuclear antibody with multiple other immunologic antibody positive (Table 1). Plain radiograph showed...
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dilated loops of small bowel with multiple air-fluid levels (Figure 1 (a)). Renal ultrasound revealed hydronephrosis, dilation of both renal pelvis, left renal pelvis dilated by 16 mm and right renal pelvis dilated by 10mm. Abdominal enhanced computed tomography (CT) revealed intestinal vascular loop-like enhancement, intestinal dilatation indicating ileus, effusion in thoracic cavity, abdominal cavity and pelvic cavity (Figure 1 (b), (c), (d)). Her anti-SSA antibody is positive. We also performed Schirmer test, BUT test, and labial gland biopsy. Results excluded the possibility of Sjogren’s syndrome.

Based on results of detailed examination, we performed a renal biopsy to confirm the diagnosis of SLE and to guide the next course of treatment. This patient’s renal pathology resulted as active focal lupus nephritis with membranous lupus nephritis (Class III-(A)+V) (Figure 2). Based on her clinical results and references [3,4, 6-8], we considered that her ileus symptoms could be related to SLE. Therefore we performed enteroscopy along with intestinal biopsy and expected to find intestinal vasculitis. However no typical vascular tissue was observed. Intestinal biopsy revealed only epithelium interstitial fibrosis, which indicated a chronic mucosal inflammation along with lymphocyte infiltration (Figure 3).

Table 1: Results of laboratory examination

| Index | ALB (g/L) | WBC (10^9/L) | Neut% | Hb (g/L) | PLT (10^9/L) | Cr (umol/L) | ANA | Anti-dsDNA | SSA-52 | SSA-60 |
|-------|-----------|--------------|-------|----------|-------------|-------------|-----|------------|--------|--------|
|       | 27.5      | 16.6         | 89.4  | 122      | 225         | 76          | 1:320| -          | 261    | 221    |

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Figure 1: Plain radiograph showed dilated loops of small bowel with multiple air-fluid levels. (b,c,d) abdominal enhanced CT revealed intestinal vascular loop-like enhancement, intestinal dilatation indicating ileus, effusion in thoracic cavity, abdominal cavity and pelvic cavity.

Figure 2: (a) Deposition of fuchsinophilic protein in mesangial, subendothelial, subepithelial space. (b) Mesangial cells and matrix lightly proliferating, glomerular basement membrane widespread thickening. (c) Focal segmental mesangial cells and matrix lightly proliferating, glomerular basement membrane widespread thickening, with a small cellular crescents, tubular epithelial vacuolar and granular degeneration, without renal interstitial and arteriole involvement. (d) Focal segmental mesangial cells and matrix lightly proliferating, glomerular basement membrane widespread thickening, with a small cellular crescents, tubular epithelial vacuolar and granular degeneration, without renal interstitial and arteriole involvement.

Figure 3: Intestinal biopsy revealed epithelium interstitial fibrosis indicating chronic mucosal inflammation along with lymphocyte infiltration.
She was given gastrointestinal decompression, colo-clyster, along with other supportive treatment including rehydration, electrolyte repletion, and nutritional supplementation. Corticosteroid therapy with methylprednisolone was used and remarkable clinical improvements were observed. Her bowel sound was heard again after 5 days, venting after 7 days, independent defecation after 10 days. Her symptoms of ileus were gradually improved. Her gastrointestinal symptoms disappeared by the time we performed intestinal biopsy.

Ethical approval: The research related to human use has been complied with all the relevant national regulations, institutional policies and in accordance the tenets of the Helsinki Declaration, and has been approved by the authors' institutional review board or equivalent committee.

Informed consent: Informed consent has been obtained from patient included in this study.

3 Discussion

SLE involves multiple systems, and the prevalence of gastrointestinal manifestations is reported by 42.5% [7]. When GI symptoms are similar to initial manifestation of SLE, there tend to be a delayed diagnosis. As an unusual complication of SLE, the diagnosis of CIPO is mainly depends on clinical symptoms and confirmed by endoscopic or radiological exclusion of mechanical causes as well as evidence of air-fluid levels in distended bowel loops. Symptoms of CIPO related to SLE such as abdominal pain and distention are non-specific. So it gives us more challenge to identify this condition.

In this report, the patient with recurrent ileus fulfills less than 4 items of the ARC criteria. No typical clinical manifestations are observed, such as malar rash. Renal biopsy results indicate lupus nephritis (active focal lupus nephritis with membranous lupus nephritis (Class III-(A) +V)). We expect to find ischemic or vasculitic damage of intestine. Unfortunately, the result of intestinal biopsy doesn't confirm our expectations. The major influential factor could be that the corticosteroid treatment had been performed for 20 days before intestinal biopsy, and the vasculitic damage may disappear with the improvement of her symptoms. According to past reports, the small bowel is more frequently involved by pseudo-obstruction (83%) [8]. To take tissue from sigmoid colon is limited by existent technology. It was possible that the pathological tissue cannot be taken. However, the interstitial change of intestinal epithelial tissue is observed.

CIPO may be life threatening in some cases. Lack of awareness of this disease will lead to misdiagnose and inappropriate treatment. Even invasive procedures will be used to cure patients.

4 Conclusion

Here we present a case of CIPO related to untypical inactive SLE. The patient’s symptoms were remarkably improved after corticosteroid treatment. The purpose of this case report is to decrease the rate of misdiagnosis when CIPO comes as the initial manifestation of SLE, and to avoid inappropriate plans of treatment in the early stages.

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Conflict of interest: Authors state no conflict of interest.

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