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Case Report

Systemic lupus erythematosus with intestinal pseudo-obstruction as initial manifestation in a middle-aged man: a case report and literature review

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Abstract: Systemic lupus erythematosus (SLE) is an unusual autoimmune inflammatory disease which can involve multiple systems and organs. Patients with SLE commonly show symptoms of gastrointestinal tract. However, intestinal pseudo-obstruction (IPO) in SLE is infrequent but severe, especially when it occurs in patients without any other manifestations. Up to now, cases of SLE with IPO as initial manifestation are extremely rare. This paper reports an additional case of a 39-year-old man in whom the first manifestations were abdominal pain, abdominal distention and vomiting. Computer tomography (CT) scan of the abdomen showed that his small bowel walls were getting thick along with inflammatory exudation. The tentative diagnosis was intestinal obstruction. He underwent surgical treatment because of the poor efficacy of conservative treatment. Nevertheless, he had a continued fever after the operation. Finally, immunologic examination proved that the diagnosis was SLE-related IPO. The patient was treated with high doses of steroids and had a good remission. SLE with IPO is very easy to be misdiagnosed and could even threaten the patient's life, so that early diagnosis and appropriate treatment are rather important.

Keywords: Systemic lupus erythematosus, intestinal pseudo-obstruction, gastrointestinal manifestations, diagnosis

Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease caused by multiple immune disorders, which is more common in young women. SLE has a variety of clinical symptoms, such as dental ulcer, arthritis, pericarditis, nephritis, anaemia, dementia and so forth, involving almost all systems and organs of the body [1, 2]. Intestinal pseudo-obstruction (IPO) is a serious complication of SLE, the mechanism may be related to the inflammations of bowel walls and mesenteries. Therefore, SLE with IPO might be a signal which suggests that the disease is active [3]. Using steroids and immunosuppressants is an effective measure for the treatment of SLE-related IPO. In general, surgical intervention is unnecessary and it dose not seem to be helpful to the sufferers [4]. As described by Xu *et al.*, there were less than 40 cases of IPO secondary to SLE which were reported in the English literature until 2014 [5]. While it was even more infrequent that intestinal obstruction appeared as the primary manifestation. We describe the case of a middle-aged man with IPO as initial symptom in the present study. Besides, we discuss the main clinical characteristics, diagnostic difficulties, treatment options and prognosis of the case with a review of the literature.

Case report

In November 29, 2011, a 39-year-old man was hospitalized in the Department of Gastroenterology of Affiliated Wujiang Hospital of Nantong University (Suzhou, China), complaining of abdominal pain and abdominal distention for one week as well as vomiting for 24 h. The patient denied any past history of rheumatism or immunopathy, and he did not present with butterfly erythema, lipsotrichia, myalgia, dental

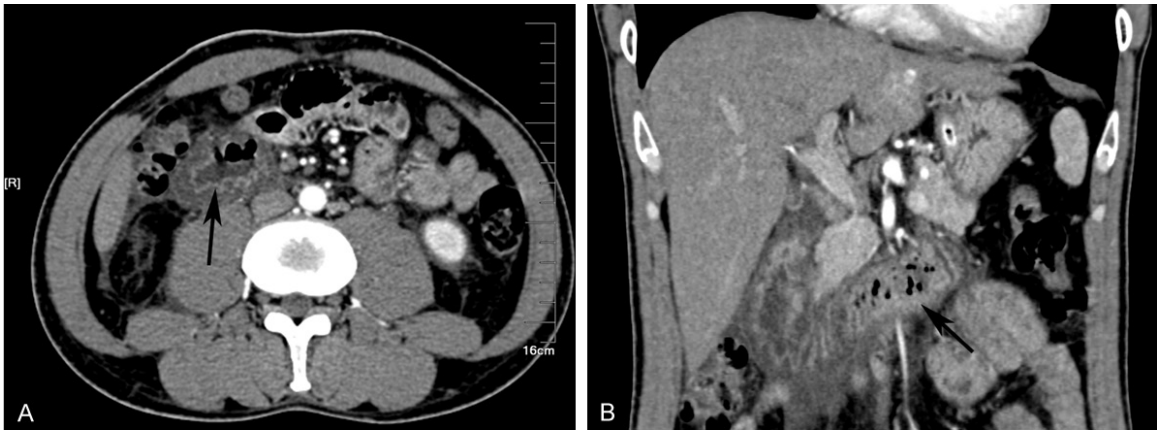

Figure 1. Computer tomography scans of the abdominal cavity, demonstrating the small bowel walls of the patient were getting thick along with inflammatory exudation (arrow). A: Coronal plane, B: Cross section.

ulcer or other SLE-associated symptoms. Physical examination showed abdominal tenderness and weak intestinal gurgling sound. Computer tomography (CT) imaging demonstrated that the small bowel walls of the patient were getting thick along with inflammatory exudation, and there was a little effusion in the abdominal cavity (**Figure 1**). The main abnormal laboratory analysis results were as follows: Albumin (Alb), 2.7 g/dl (normal range, 3.5-5.5 g/dl); hemoglobin (Hb), 10.8 g/dl (normal range, 12.0-16.0 g/dl); erythrocyte sedimentation rate (ESR), 48 mm/h (normal range, <15/h); C-reactive protein (CRP), 15.9 mg/l (normal range, <10 mg/l). Other completed examinations and tests were unremarkble. The tentative diagnosis was intestinal obstruction.

The patient received conservative treatment including fasting, gastrointestinal decompression, inhibition of gastric acid secretion, anti-inflammatory therapy, infusion of albumin, correction of water-electrolyte imbalance, nutritional support and so on. Yet the patient's condition took a turn for the worse. For further diagnosis and treatment, he transferred to the Department of Gastroenterology of The First Affiliated Hospital of Soochow University (Suzhou, China) in December 15, 2011. Shortly afterward, he underwent exploratory laparotomy on account of his abdominal symptoms were not relieved. The surgeons found that the patient's jejunum and ileum appeared dull-red and the bowel walls were edematous, with a large quantity of effusion in his abdominal cavity. Meanwhile, they noticed that the duodenum was clearly thickening and the enteric cavity was obstructed. Consequently, he received gastrojejunostomy in order to alleviate the suffering. Unfortunately, the effect of the operation was not ideal. The patient presented a continuous fever with wandering joint pain for more than a month. Then further examinations and tests were taken, hydrothorax and seroperitoneum were detected by ultrasonic inspection, as well as obvious albuminuria was found by uronoscopy. Considering that the patient had multi-systemic lesions, immunological analyses were finished. The results revealed that antinuclear antibody, anti-double-strand DNA antibody and anti-Sm antibody were positive. According to the SLE classification standards recommended by American College of Rheumatology (ACR) in 1997, the patient was diagnosed with SLE-related IPO.

Since the definitive diagnosis was established, the patient was treated with methylprednisolone (1 g/day) for 3 days and his abdominal symptoms were significantly improved. We continued to treat him by using prednisone (1.5 mg/kg/day). Until April 6, 2012, the does of prednisone had been reduced to 10 mg/d gradually and the patient was discharged. At the time of writing this paper, the patient's condition is still well.

Discussion

SLE is a multi-systemic autoimmune disease with unclear etiology [6]. It occurs mostly in the person aged between of 20 to 40, and women are far more likely to fall victim to SLE. At times, symptoms of SLE may be relieved or completely

disappeared without treatment in some patients. In recent years, atypical cases of SLE are increasing gradually. Digestive disorders in patients with SLE are less common than the other major system or organ involvements [7]. IPO has been recognized as a rare and severe complication of SLE. It is characterized by ineffective intestinal propulsion and can be expressed as abdominal pain, abdominal distention, constipation or vomiting, but there is no mechanical obstruction in the digestive tract [8-10]. IPO generally occurs when the disease is active, but as described by Leonardi *et al.*, it could also be found in inactive SLE sometimes [3]. It is rarely seen that IPO manifesting itself as the primary condition of SLE. Under these circumstances, the clinical features are very similar to those of simple gastrointestinal disease. Hence, patients of SLE with IPO as initial symptom are extremely easy to be misdiagnosed. A recent study by Wang *et al.* has found that 16 of 40 cases (40%) had gastrointestinal symptoms as the initial manifestation of SLE, and 11 cases (69%) of them were misdiagnosed [4].

The patient in the present study was a 39-year-old man. In the early stage of the disease, he only showed the clinical presentations of abdominal pain, abdominal distention, vomiting, hypoproteinemia and mild anemia, which did not conform to the criteria for SLE. With the further development of the disease, he began to exhibit symptoms of multiple systems involved, including fever, wandering joint pain, polyserositis and albuminuria, which led to the diagnosis of SLE. So we ought to pay more attention to the digestive manifestations of patients with SLE and take necessary investigations in order to make a clear diagnosis as soon as possible.

CT examinations are probably most useful for the diagnosis of SLE-related IPO. As demonstrated in the previous studies, abdominal CT scans of the patients with IPO can usually reveal the characteristic imaging features of this disease, such as target dilated loops, thickened bowel walls, multiple fluid levels, seroperitoneum, urinary system involvement and so on [11-15]. Chen *et al.* found that enteroscopy was needed to be performed after radiologic examinations, and by doing so, other causes such as ischemic bowel, Crohn's disease, or small-bowel lymphoma may be excluded [6]. Immunologic tests can also play an important role in the diagnostic procedure of IPO. Mok *et al.* reported 6 patients with SLE-related IPO and showed that 5 cases (83.3%) of them had anti-Ro antibodies, 4 cases (66.7%) of them had anti-RNP antibodies [16]. In our patient, antinuclear antibody, anti-double-strand DNA antibody and anti-Sm antibody were positive.

The usual treatment for SLE-related IPO is to apply high doses of steroids, and immunosuppressants are often used in combination with steroids. Most patients have a good response to these drugs [17-19]. However, the effect of surgical intervention is poor. Wang *et al.* reported that 11 patients with IPO received surgical management, and all of them relapsed after surgery [4].

Conclusion

IPO as initial manifestation in SLE patients is extremely rare, but it is a severe situation that can potentially be life-threatening. Early recognition and appropriate treatment are rather important that can lead to a better prognosis and avoid unnecessary surgical intervention.

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Disclosure of conflict of interest

None.

Abbreviations

SLE, Systemic lupus erythematosus; IPO, intestinal pseudo-obstruction; CT, Computer tomography.

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