

Case Report

Gastrointestinal symptom due to lupus peritonitis: a rare form of onset of SLE

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Abstract: Serositis is commonly seen in systemic lupus erythematosus (SLE). Approximately 16% of patients with SLE have pleural or pericardial involvement. However, peritoneal involvement is extremely rare, and SLE with ascites as the first manifestation is an even rarer condition. This is the case report of a 19-year old male with discoid lupus who evolved with gastrointestinal symptoms as the first manifestation of the disease, characterized by significant abdominal distension and pain, asthenia, vomiting, and signs of ascites. An abdominal CT scan demonstrated ascites and marked edematous thickening of the bowel wall, which appeared as “target sign”, and “double-track sign”. Laboratory tests showed that his serum complement levels decreased and that he was positive for anti-nRNP/Sm antibodies, anti-Sm antibodies, anti-SS-A antibody, and anti-nuclear antibodies. The patient was treated with prednisone and chloroquine, with substantial improvement of his condition.

Keywords: Systemic lupus erythematosus, serositis, ascites, lupus peritonitis, CT

Introduction

Systemic lupus erythematosus (SLE) is a chronic multisystem autoimmune disease with a wide spectrum of potentially serious symptoms, characterized by autoantibodies against nuclear antigens and deposition of immune complexes in several tissues [1]. The incidence of disease is 10-fold higher among females compared to males, and incidence peaked in the population aged 25-44 [2]. Serositis is a common finding among the wide range of manifestations of SLE patients. Approximately 16% of SLE patients have pleuritis and/or pericarditis, but peritoneal involvement is extremely rare, and SLE with ascites as the first manifestation is an even rarer condition [1].

This is the report of a patient initially diagnosed with discoid lupus who evolved with systemic manifestations, such as chronic peritoneal serositis and colitis.

Case report

The patient is a 19-year-old Chinese male, complaining of progressive increase of his abdomi-

nal pain associated with nausea and vomiting for three days. Overall, he had been well until three days before his presentation. He denied any pattern of pain irradiation, association with food, dysuria or fever. He also reported the defecation 1-3 times per day. He denied alcoholism and illicit drug use, and smoking.

On physical exam, his general state of health was regular, acyanotic, anicteric, febrile (37.8°C), eupneic, and thin. His blood pressure was 120/70 mmHg, and pulse rate was 92 beats/min. His throat, neck, and lungs were normal, and no heart murmur or abnormal heart sounds were audible. No lymph nodes were palpable. His abdomen was distended, and diffusely tender, and the upper abdomen and periumbilical area was tenderness, with neither muscle guarding nor rebound tenderness. No masses and collateral circulation were palpable. Signs of ascites were positive. Liver and spleen were non-palpable. Digital rectal examination did not reveal feces, blood, or mass in the rectal ampulla. Mild, cold, painless, pitting edema was observed in both lower limbs. The osteoarticular system showed normal.



Figure 1. Abdominal CT scan showing dilated bowel, focal or diffuse bowel wall thickening, abnormal bowel wall enhancement (target sign) mesenteric oedema, engorged mesenteric vessels, and ascites.

The previous laboratory tests of patient were as follow: blood routine tests remained WBC $3.3 \times 10^9/L$, NE% 75.11%; urine routines remained PRO (2+)/KET (3+), abdominal ultrasound revealed only mild ascites (18 mm deep), after three days, the reexamination of abdominal ultrasound revealed moderate ascites (84 mm deep). On admission, his initial laboratory tests were as follows: blood routine tests remained WBC $2.5 \times 10^9/L$, NE% 71.9%. Serologies for HIV, viral hepatitis, and VDRL were negative. ALT, ASG, GGT, and electrolytes levels were normal. LDH, 267 μ/l , and serum albumin: 33.7 g/L; BUN and creatinine was normal; 24-h proteinuria: 0.51 mg/24 h; Fresh stool test and stool culture showed no abnormalities. Urinalysis evidenced erythrocytes 19/ μl , WBC 105/ μl , and PRO (2+)/KET (3+). Serologies for C-reactive protein (CRP) was slight elevation (10.88 mg/L), and erythrocyte sedimentation rate (ESR): 33 mm/h. CEA, CA199 and AFP levels were normal.

Puncture of the ascitic fluid showed the following: total leukocyte count of $1266 \times 10^6/L$ (neutrophils cells, eosinophils inflammatory cells); frequent mesothelial cells; LDH, 514 U/L; TP (total protein), 41.3 g/l, and Rivalta, positive. The following tests were negative: LE cells; bacterioscopy; Koch bacillus; CEA; ADA; and oncotic cytology.

Plain chest X-ray showed normal. Abdominal CT confirmed the ultrasound findings and showed intestinal wall oedema and circumferential wall thickening and target sign in small and large bowels (Figure 1).

Based on the findings, eosinophilic enteritis was taken into account. In order to rule out rheumatoid immune system disease, the auto-immune antibodies, immune function, and 24 hours urinary protein quantitative were checked. The patient was temporarily treated with gastrointestinal decompression, anti-infection, fluid infusion, inhibition of gastric acid secretion, somatostatin analogues maintenance and nutritional support treatment.

After five days of treatment, his clinic symptom was improved, but still had abdominal pain, abdominal distension, and drainage of the much bile-like liquid by nasogastric suction. And the reexamination of ultrasound revealed moderate abdominal ascites (88 mm deep) and right-sided hydrothorax (25 mm deep).

At this moment, the relevant test results were come out: complement levels were reduced C3 0.55 mg/ml (normal 0.7-2 mg/ml); C4 0.1 mg/ml (normal 0.2-0.5 mg/ml); anti-SSA antibody, antinuclear antibodies (ANA) and anti-dsDNA antibody were positive. The patient was non-reactive for the following antibodies: anti-La; anti-cardiolipin; lupus anticoagulant; anti-SM; anti-RNP; anti-SCL-70; and anticentromere. Coombs test was normal.

Systemic lupus erythematosus was suspected based on the American College of Rheumatology criteria: malar rash; discoid lupus; photosensitivity; persistent proteinuria > 500 mg/dL; serositis; and ANA [3]. Prednisone (60 mg/day) was introduced, and chloroquine (250 mg/day) was maintained. The patient showed improvement of his general state of health, a reduction in abdominal circumference, and pleural effusion and ascites reduce, and initiated outpatient clinic follow-up.

Three months after beginning treatment, the patient has recovered well. He is currently followed up at the outpatient clinic, remains asymptomatic, with no ascites, and has regular intestinal rhythm (once or twice a day) with soft stools and no bleeding.

Discussion

Systemic lupus erythematosus (SLE) is a multifactorial autoimmune disease that mostly affects young women [2]. Inflammation of the pleural and pericardial serous membranes, although non-specific, is relatively common, however, peritoneal serositis with ascites is extremely rare [1]. SLE can affect the entire gastrointestinal tract [2, 3]. However, the development of gastrointestinal complications in patients with SLE is not related to drugs and infections are rare [4].

Symptoms related to the disease itself are not as common as the involvement of other organs, such as in lupus nephritis. On the other hand, the incidence of gastrointestinal manifestations can be clinically underestimated, since some are non-specific and abdominal symptoms can be absent [5]. Abdominal pain and ascites is a common clinical manifestation of several diseases. It is a real challenge that differentiating a patient with an acute abdomen secondary to SLE as the sole presenting symptom.

SLE-related gastrointestinal involvement is clinically important, and the causes to be considered include mesenteric vasculitis, protein-losing enteropathy, intestinal pseudo-obstruction, acute pancreatitis and other rare complications such as celiac disease, inflammatory bowel diseases [4]. And, the intestinal involvement might have been secondary to mesenteric vasculitis. The hypoproteinemia of the patient could be justified by both proteinuria and protein-losing enteropathy. CT is the important diagnostic tool for SLE. Common CT findings in SLE include dilated bowel, focal or diffuse bowel wall thickening, abnormal bowel wall enhancement (target sign) mesenteric oedema, engorged mesenteric vessels, and ascites [6]. And the CT is helpful to differentiate SLE from thromboembolic disease, whereby the bowel wall thickening tends to be restricted to vascular territories. However, the lack of specificity of the 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 [7].

In the present case, the patient had abdominal pain nausea and vomiting associated with mild

ascites (18 mm deep) as his initial SLE manifestation. In clinical practice, malignant ascites and tuberculous ascites were relatively common. When facing a patient with visible ascites, the first essential step is to correctly diagnose the cause of the ascites. Ascites can be acute or chronic, with or without pain, and two factors could be implicated: portal hypertension or peritoneal diseases. Specific diagnostic approach is initiated by the distinction between those two triggering factors by use of diagnostic paracentesis. Ascitic fluid characteristics from lupus peritonitis includes a Serum Ascites Albumin Gradient (SAAG) of less than 1.1 and can include a wide range of WBCs from 10 to 1630/mm³, and a range of fluid protein from 34 to 47 mg/L [8].

Lupus peritonitis has been reported in the literature as a major cause of ascites in lupus patients. But it should be considered an exclusion diagnosis, demanding an extensive investigation for the most common causes of exudative ascites, such as peritoneal carcinomatosis, primary mesothelioma, peritoneal pseudomyxoma, hepatocellular carcinoma, peritoneal tuberculosis, nephrotic syndrome, protein-losing enteropathy, and severe malnutrition [8].

The case of lupus ascites here is different from previous ones reported in the literature. The patient was young man, who evolved with gastrointestinal symptoms as the first manifestation of the disease. The ascites developed over three to two weeks, was not associated with rash, or arthritis. Heart, liver, and kidney disease were ruled out and there was no obstruction of the inferior vena cava or mesenteric vessels. However, Puncture of the ascitic fluid showed and abdominal CT scan suggested the diagnosis of Lupus peritonitis.

The prognosis of SLE peritonitis is usually good, and current therapeutic schedules are based on non-steroidal anti-inflammatory drugs and corticosteroids. But the refractory cases still had be reported. In these cases, it could be necessary that immunomodulators or immunosuppressors, as well as surgical procedures [9].

Conclusion

Lupus peritonitis as the initial SLE manifestation is rare, which is most likely due to its clinical underestimation. Even though, lupus perito-

nititis should be considered an exclusion diagnosis, requiring extensive clinical evaluation in search for alternative causes of exudative ascites. CT is the important diagnostic tool for lupus peritonitis. Prognosis is usually good, and treatment is based on the use of non-steroidal anti-inflammatory drugs and corticosteroids, with good response. For refractory cases, individualized alternative measures are indicated.

Disclosure of conflict of interest

None.

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References

- [1] Pott Junior H, Amate Neto A, Teixeira MA and Provenza JR. Ascites due to lupus peritonitis: a rare form of onset of systemic lupus erythematosus. *Rev Bras Reumatol* 2012; 52: 116-119.
- [2] Somers EC, Marder W, Cagnoli P, Lewis EE, DeGuire P, Gordon C, Helmick CG, Wang L, Wing JJ, Dhar JP, Leisen J, Shaltis D and McCune WJ. Population-based incidence and prevalence of systemic lupus erythematosus: the Michigan Lupus Epidemiology and Surveillance program. *Arthritis Rheumatol* 2014; 66: 369-378.
- [3] Tan EM, Cohen AS, Fries JF, Masi AT, McShane DJ, Rothfield NF Schaller JG, Talal N, Winchester RJ. The 1982 revised criteria for the classification of systemic lupus erythematosus. *Arthritis Rheum* 1982; 25: 1271-7.
- [4] Yuan SW, Li H, Chen DY, Qiu Q, Lian F, Zhan ZP, Liang LQ, Xu HS and Yang XY. [Associated factors for the occurrence of lupus enteritis]. *Zhonghua Yi Xue Za Zhi* 2013; 93: 1162-1164.
- [5] Tian XP and Zhang XA. Gastrointestinal involvement in systemic lupus erythematosus: Insight into pathogenesis, diagnosis and treatment. *World J Gastroenterol* 2010; 16: 2971-2977.
- [6] Chng HH, Tan BE, Teh CL and Lian TY. Major gastrointestinal manifestations in lupus patients in Asia: lupus enteritis, intestinal pseudo-obstruction, and protein-losing gastroenteropathy. *Lupus* 2010; 19: 1404-1413.
- [7] Lee CK, Ahn MS, Lee EY, Shin JH, Cho YS, Ha HK, Yoo B and Moon HB. Acute abdominal pain in systemic lupus erythematosus: focus on lupus enteritis (gastrointestinal vasculitis). *Ann Rheum Dis* 2002; 61: 547-550.
- [8] Forouhar-Graff H, Dennis-Yawing K and Parke A. Insidious onset of massive painless ascites as initial manifestation of systemic lupus erythematosus. *Lupus* 2011; 20: 754-757.
- [9] Zhou QG, Yang XB, Hou FF and Zhang X. Successful treatment of massive ascites with intraperitoneal administration of a steroid in a case of systemic lupus erythematosus. *Lupus* 2009; 18: 740-742.