Case Report

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Ascites as primary manifestation of systemic diseases: two case reports.

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ABSTRACT

Ascites is rare in systemic autoimmune diseases, and being exceptionally the presenting symptom. We report two cases of systemic autoimmune diseases which presented with ascites as the first manifestation of the disease.

Keywords
Ascites; autoimmune disease; systemic lupus erythematosus; mixed connective tissue disease.

INTRODUCTION

Ascites is most commonly caused by cirrhosis. Approximately 15% of patients with ascites have non-liver causes and 5% have two or more causes of ascites (1). However, ascites is a rare manifestation of systemic autoimmune diseases. Here, we report two patients who had ascites as the presenting symptom of mixed connective tissue disease (MCTD) and systemic lupus erythematosus (SLE).

CASE REPORT

Case 1

A 27-year-old lady presented with increased abdominal girth and vomiting. Her past medical history revealed a neglected Raynaud phenomenon. Two weeks prior to presentation, she was seen by a dermatologist for bullous cutaneous rash on her chest, neck and thighs and was started on oral and topical Acyclovir. Her skin lesions resolved completely within 3 weeks. On admission, her temperature was 36.7 °C, her blood pressure was 120/80 mmHg and her heart rate was 85 beats per minute. Her physical examination revealed abdominal distension with shifting dullness consistent with massive ascites. Laboratory tests showed white blood cell (WBC) count = 8840/mm3 with left shift (neutrophils = 76%), hemoglobin = 14.7g/dl, platelets = 377000/mm3, international normalized ratio (INR) = 1.35, albumin = 3.65g/dl, C-reactive protein (CRP) = 23mg/L and erythrocyte sediment rate = 87mm. Liver function tests, blood urea nitrogen (BUN) and serum creatinine were normal. HBV and HCV serology were negative. Abdominal ultrasound confirmed the diagnosis of ascites. CT scan revealed minimal pericardial effusion, dilated esophagus with intraesophageal stasis, bilateral pleural effusion more on the right side and large amount of ascites with normal liver (Figure 1). Analysis of the ascitic fluid: WBC = 128/mm3 (lymphocytes = 97%), red blood cell (RBC) = 670/mm3, total protein = 5.23g/dl and albumin = 2.82g/dl (serum ascites-albumin gradient (SAAG) = 1.33mg/dl) with negative culture, cytology, and PCR for Mycobacterium Tuberculosis. Transthoracic echocardiogram showed minimal pericardial effusion. Esophagogastroduodenoscopy was done to evaluate the dilated esophagus found on CT scan and showed dilated esophagus, inflammatory mucosa, diffuse gastritis with food stasis. Biopsies: nonspecific duodenitis, gastritis with moderate activity, positive Helicobacter pylori and severe erosive esophagitis with presence of eosinophils. Pleural fluid studies were not conclusive as well as skin biopsy of her remaining lesions (figure 2) (Bullous dermatitis). An exploratory laparoscopy was done and biopsies from the peritoneum, the liver and the omentum showed leukocytoclastic vasculitis with non-monoclonal immune deposits. A complementary immunologic panel was positive for ANA and anti-RNPwith low complement levels (Table 1).

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Table 1 : immunologic panel of the patient's serum

Test	Result
ANA	Positive 1:640
Anti-DNA	Negative
Anti-SSA	Negative
Anti-SSB	Negative
Anti-RNP	Positive
Anti-Sm	Negative
Anti-Scl-70	Negative
Anti-Jo1	Negative
p-ANCA	Negative
c-ANCA	Negative
RF	Negative
C3	Low (5.5)
C4	Low (5.8)
IgA	232 mg/dl
IgM	178 mg/dl
IgG	829 mg/dl
Cold agglutinin	Negative

Figure 1

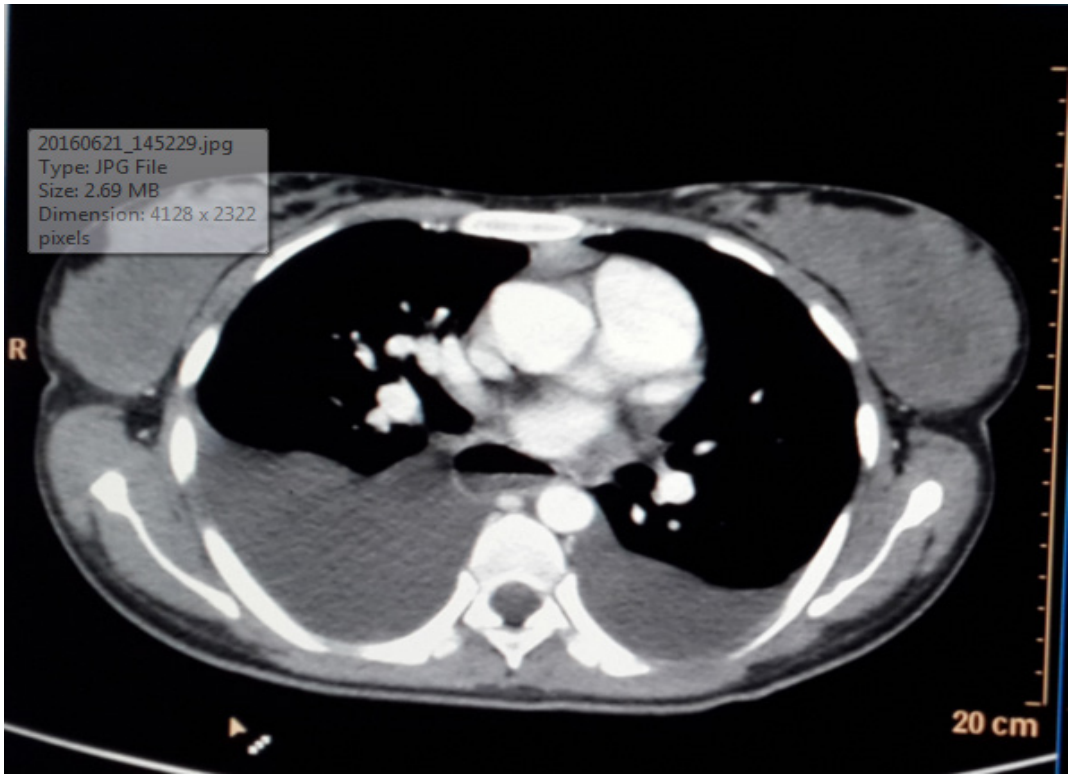

Figure 1: CT scan showing bilateral pleural effusion and dilated esophagus with intraesophageal stasis

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Figure 2

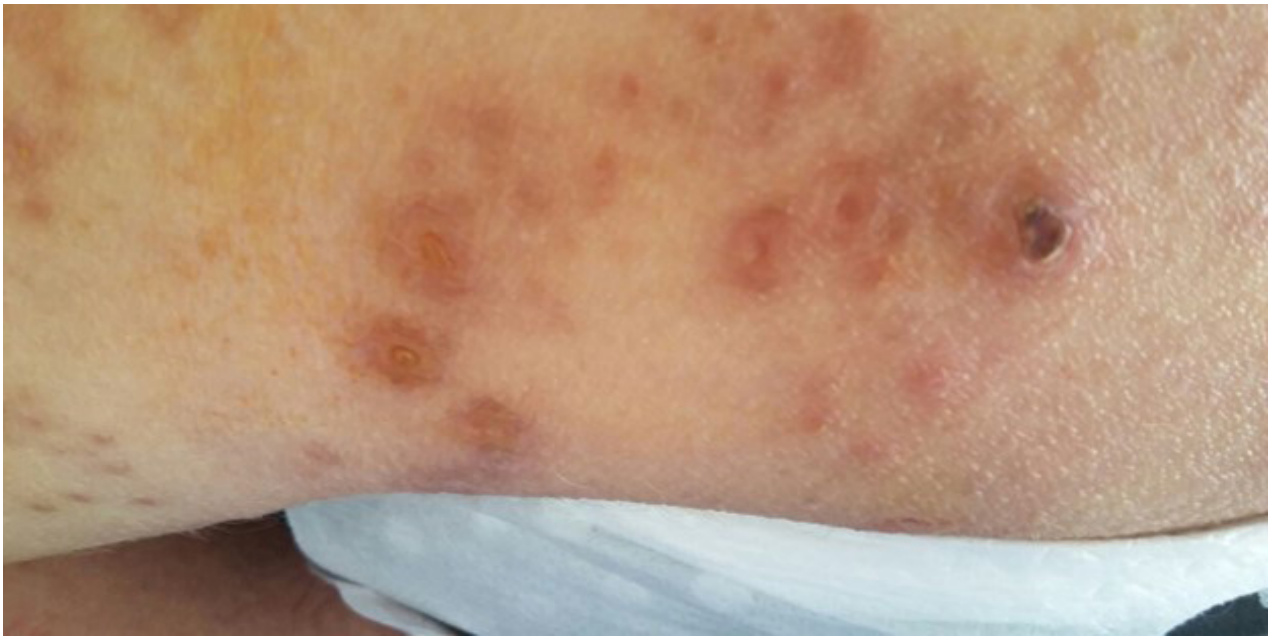

Figure 2 : Patient's residual skin lesions.

Our patient had Raynaud's phenomenon and systemic involvement of the lungs, heart, skin, esophagus and peritoneum in conjunction with positive RNP antibodies. All of these findings make the diagnosis of MCTD. She received Mycophenolic acid along with an induction therapy with steroid that was tapered and stopped later on with complete resolution of her ascites.

Case 2

A 30-year-old lady, previously healthy, presented three months' post-partum because of increase abdominal girth, decrease appetite, weight loss and watery, non-mucoid, non-bloody diarrhea. On presentation, the patient was afebrile and hemodynamically stable. On physical examination, she had abdominal distension, dullness to percussion with shifting dullness. Heart and lung examinations were normal. Laboratory studies including WBC count, serum creatinine, electrolytes, liver function tests and CRP were normal. Urine analysis showed 3+ proteinuria with 15-20 RBC/hpf and 24-hour urine collection contained 1.1g of proteins. An abdominal ultrasound showed abundant ascites. Drainage of the ascitic fluid was done and analysis showed WBC = 150/mm3 (lymphocytes = 80%), RBC = 162/mm3, albumin = 2.3 mg/dl (SAAG = 0.7mg/dl). Culture and cytology of the ascitic fluid were negative. A porto-mesenteric vein thrombosis was ruled out by negative abdominal Doppler ultrasonography.

A systemic disease was suspected, so an auto-immune profile was conducted and revealed an elevated anti-nuclear antibody (ANA) titer (>1/1280) with highly positive Anti-DNA (>800 UI/ml). She had also positive Anti-SSA (105 UI/ml), Anti-Sm (32 UI/ml) and Anti-RNP (42 UI/ml) titers with low complement levels (C3 = <30mg/dl and C4 = 7.5mg/dl). Anti-SSB, Anti-Jo1, Anti-Scl 70 and Rheumatoid factor were negative.

Based on the American College of Rheumatology, this patient had 4 of the 11 criteria for the diagnosis of SLE: serositis (peritonitis with ascites), renal involvement (proteinuria = 1.1g/day), high titer of ANA (>1:1160) and positive anti-DNA and anti-Sm antibodies. She responded well to Azathioprin (Imuran) which was discontinued by the patient was lost to follow up.

DISCUSSION

Cirrhosis is the most common cause of ascites accounting for 85% of cases (1). Approximately 15% of patient with ascites have etiologies not related to the liver, and 5% have two or more causes (1). Ascites is rarely caused by systemic autoimmune diseases. We report here one of two cases of MCTD and one of six cases of SLE who presented with ascites as a first manifestation of the disease.

MCTD is an overlap syndrome that includes the clinical characteristics of SLE, systemic sclerosis and polymyositis, in the setting of high titers of anti-U1 ribonucleoprotein (RNP) antibodies (2). MCTD can affect any organ but gastrointestinal manifestations are rare. Heartburn and dysphagia are the most common symptoms and to a lesser extent bowel perforation and malabsorption (2). There are reported cases of pneumatosis intestinalis, chronic active hepatitis and autoimmune hepatitis (3-6), and one case of ascites as a first presentation of MCTD (7).

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SLE is a chronic inflammatory autoimmune disease that can affect any organ. It can affect the GI tract causing GI vasculitis, esophageal dysmotility, Pneumatosis cystoides intestinalis, intestinal pseudoobstruction, malabsorption, protein-losing enteropathy, acute pancreatitis and acute peritonitis with ascites (8). The latter occurs rarely in SLE especially as a first presentation of the disease. To our knowledge, only seven cases were reported in the literature of ascites as a first presentation of SLE (9-15). The pathophysiology of ascites in SLE can be multifactorial. Three factors have been suggested: auto-reactivation of B lymphocytes producing autoantibodies that bind to corresponding antigen forming immune complexes that deposit on the peritoneum and trigger a local inflammatory reaction (10), vasculitis of the peritoneal vessels or the serous membrane of abdominal organs that may be the underlying mechanism (10) and antiphospholipid antibodies that accentuate chronic ascites in some patients (11).

CONCLUSION

The GI manifestations of systemic diseases are rare and non-specific, they are most commonly attributed to drug side effects. It is important to make the right diagnosis as some manifestations can be life-threatening. The clue is to combine clinical manifestations with appropriate laboratory tests and investigations. We report here two cases of systemic diseases which presented with ascites as initial symptom.

REFERENCES

1. Runyon BA, Aasld. Introduction to the revised American Association for the Study of Liver Diseases Practice Guideline management of adult patients with ascites due to cirrhosis 2012. Hepatology. 2013;57(4):1651-3.

2. Marshall JB, Kretschmar JM, Gerhardt DC, Winship DH, Winn D, Treadwell EL, et al. Gastrointestinal manifestations of mixed connective tissue disease. Gastroenterology. 1990;98(5 Pt 1):1232-8.

3. Lynn JT, Gossen G, Miller A, Russell IJ. Pneumatosis intestinalis in mixed connective tissue disease: two case reports and literature review. Arthritis Rheum. 1984;27(10):1186-9.

4. Rolny P, Goobar J, Zettergren L. HBsAg-negative chronic active hepatitis and mixed connective tissue disease syndrome. An unusual association observed in two patients. Acta Med Scand. 1984;215(4):391-5.

5. Tomsic M, Ferlan-Marolt V, Kveder T, Hojker S, Rozman B. Mixed connective tissue disease associated with autoimmune hepatitis and thyroiditis. Ann Rheum Dis. 1992;51(4):544-6.

6. Fernandes C, Bungay P, O'Driscoll BR, Herrick AL. Mixed connective tissue disease presenting with pneumonitis and pneumatosis intestinalis. Arthritis Rheum. 2000;43(3):704-7.

7. Linana Santafe JJ, Calvo Catala J, Hortelano Martinez E, Roig Lopez de los Mozos A, Gonzalez-Cruz Cervellera I, Herrera Ballester A. [Acute ascites as the onset of mixed connective tissue disease]. An Med Interna. 1993;10(10):504-6.

8. Ebert EC, Hagspiel KD. Gastrointestinal and hepatic manifestations of systemic lupus erythematosus. J Clin Gastroenterol. 2011;45(5):436-41.

9. Hammami S, Bdioui F, Ouaz A, Loghmari H, Mahjoub S, Saffar H. Successful treatment of massive ascites due to lupus peritonitis with hydroxychloroquine in old- onset lupus erythematosus. Pan Afr Med J. 2014;18:165.

10. Pott Junior H, Amate Neto A, Teixeira MA, Provenza JR. Ascites due to lupus peritonitis: a rare form of onset of systemic lupus erythematosus. Rev Bras Reumatol. 2012;52(1):116-9.

11. Corbella X, Mitjavila F, Campoy E, Saez A, Moga I, Vidaller A. Chronic ascites in late onset systemic lupus erythematosus with antiphospholipid antibodies. J Rheumatol. 1994;21(6):1141-3.

12. Ito H, Nanamiya W, Kuroda N, Inoue M, Sasaoka A, Chijiwa T, et al. Chronic lupus peritonitis with massive ascites at elderly onset: case report and review of the literature. Intern Med. 2002;41(11):1056-61.

13. Weinstein PJ, Noyer CM. Rapid onset of massive ascites as the initial presentation of systemic lupus erythematosus. Am J Gastroenterol. 2000;95(1):302-3.

14. Nakamura K, Sato Y, Kawagoe K, Asaba N, Hirata M, Tokunaga K, et al. [Elderly case of systemic lupus erythematosus who showed lupus peritonitis as the initial symptom]. Nihon Shokakibyo Gakkai Zasshi. 2007;104(10):1486-91.

15. Aljebreen AM, Al-Mogairen SA. Refractory ascites as the first presentation of systemic lupus erythematosus. Arab J Gastroenterol. 2014;15(3-4):161-2.