

77283\_Auto\_Edited.docx

**Name of Journal:** *World Journal of Clinical Cases*

**Manuscript NO:** 77283

**Manuscript Type:** CASE REPORT

**Systemic lupus erythematosus presenting with progressive massive ascites and CA-125 elevation, Tjalma syndrome? A case report**

Wang JD *et al.* Tjalma syndrome

Jun-Di Wang, Yan-Fei Yang, Xian-Feng Zhang, Jiao Huang

## Abstract

### BACKGROUND

Ascites, pleural effusion and raised CA-125 in the absence of malignancy in systemic lupus erythematosus (SLE) is known as pseudo-pseudo Meigs' syndrome (PPMS).

### CASE SUMMARY

We report a special case of a SLE patient presenting with PPMS. She presented with ascites and elevated CA-125 in the absence of benign or malignant ovarian tumor and no pleural effusions, which is an unusual presentation for this rare condition.

### CONCLUSION

Tjalma syndrome can present with massive ascites alone without pleural or pericardial effusions.

**Key Words:** Tjalma syndrome, PPMS, SLE, Ascites , CA-125, Case report

wang J, Yang Y, Zhang X, huang J. Systemic lupus erythematosus presenting with progressive massive ascites and CA-125 elevation, Tjalma syndrome? <sup>3</sup> A case report and literature review. *World J Clin Cases* 2022; In press

**Core Tip:** We report a special case of a SLE patient presenting with PPMS. She presented with ascites and elevated CA-125 in the absence of benign or malignant ovarian tumor and no pleural effusions, which is an unusual presentation for this rare condition : Tjalma syndrome can present with massive ascites alone without pleural or pericardial effusions.

## INTRODUCTION

Systemic lupus erythematosus is a chronic, autoimmune disease with multiple-systemic disorders. Tjalma syndrome, also known as PPMS, is a clinical manifestation of SLE that

is characterized by ascites, pleural effusions and elevated CA-125 in the absence of benign or malignant ovarian tumor<sup>1</sup>. Massive ascites are rare in SLE patients without any other complications. Herein we report a special case of a SLE patient presenting with PPMS. She presented with ascites and elevated CA-125 but no pleural effusions.

## **CASE PRESENTATION**

### ***Chief complaints***

A 23-year-old woman presenting with nausea, vomiting and distention for 2 wk without abdominal pain, diarrhea, rashes or arthralgia.

### ***History of present illness***

The patient had presented herself to an outside hospital 10 days ago where physical examination revealed distended abdominal wall while abdominal computed tomography (CT) scan revealed massive ascites (Figure 1). Laboratory examinations at the outside hospital showed markedly elevated CA-125 at 1685 U/mL (0-35U/mL). Ascitic fluid analyses revealed negative results from Rivalta tests. After diuresis treatment for 7 days, the amount of ascites in the patient was gradually reduced. However, there were no changes in nausea or vomiting.

**1**

### ***History of past illness***

The patient had a history of immune thrombocytopenia for two years and was administered with a long-term maintenance dose of 5 mg/d prednisone.

**1**

### ***Personal and family history***

The patient denied any family history.

### ***Physical examination***

Patient temperature and blood pressure were 37.2°C and 123/82 mmHg, respectively, whereas her heart and respiratory rates were 89 beats/min and 20 breaths/min,

respectively. No rales were heard in lung auscultation, and her heart-beat was regular without murmurs. Her abdomen was distended, shifting dullness was positive and neither her liver nor spleen were palpable. Physical examination of other parameters did not reveal any abnormalities.

### *Laboratory examinations*

White blood cell (WBC) counts  $6.8 \times 10^9/L$ , Neutrophil % (N%) 77.9%, Hemoglobin (Hb) 100 g/L, Platelet (PLT)  $130 \times 10^9/L$ , Total protein (TP) 58 g/L, Albumin (ALB) 31.6 g/L, D- dimer 1910  $\mu g/L$ , and Ferritin 37.7  $\mu g/L$ . The 24 h urine protein quantitate was 74 mg/24 h. Antinuclear antibody (ANA) 1:100 (+), Anti-Sjogren's syndrome A antigen antibody (anti-SSA) (+), C3 0.46 g/L (0.79–1.52), and C4 0.11 g/L (0.12–0.36). Lymphocyte subset findings were: Total lymphocytes  $500 \times 10^6/L$ , T-cell lymphocytes (CD3)  $226 \times 10^6/L$ , B-cell lymphocytes (CD19)  $265.9 \times 10^6/L$ , T-helper lymphocytes (CD4)  $54.4 \times 10^6/L$ , NK-cells (CD16/56)  $5.6 \times 10^6/L$ , and CD4/CD8 0.71. In addition, the tumor marker (CA125) was 439.9 U/mL, whereas other tumor markers, including AFP, CEA, NSE, CA153, CA199, and  $\beta$ -HCG were normal. Moreover, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), ANCA, index of autoimmune liver diseases, IgG4, HBsAg and HIV were all found to be within normal ranges. T-SPOT showed negative results.

### *Imaging examinations*

Small bowel enhanced CT revealed a swollen gastric wall. Small bowel wall and colon wall were slightly thickened with abnormal bowel enhancement. The number of mesenteric vessels was increased and mesenteric vessels were engorged exhibiting a “comb sign” appearance (Figure 2). Enhanced MRI scans of the pelvic tumor showed bilateral ovaries with enlarged multifocal cystic lesions, thus, endometriotic cysts were considered. Abdominal ultrasound showed abdominal effusions while portal ultrasound observations were normal. Ultrasonic examinations did not reveal any

pleural or pericardial effusions. Gastrointestinal endoscopy revealed diffuse edema of gastric and colon walls.

### **FINAL DIAGNOSIS**

Tjalma syndrome, Protein-losing enteropathy, Lupus cystitis, Clostridium difficile infection

### **TREATMENT**

20 mg intravenous methylprednisolone and 0.2 g hydroxychloroquine per day.

### **OUTCOME AND FOLLOW-UP**

There was a subsequent improvement in nausea and vomiting during her hospital stay while her ascites were reduced. However, she later presented with violent vomiting, and 7 days after admission, she was vomiting moderate amounts of a coffee-like liquid. Then, the patient started presenting with yellow watery diarrhea. Ultrasonographic examinations and abdominal CT scans showed bilateral hydronephrosis and hydroureter in addition to bladder wall thickening, and small abdominal effusions. The fecal occult blood test was positive and stool cultures revealed an infection of Clostridium difficile. Tests for Clostridium difficile toxins A and B were positive. Then, she was treated with 80 mg intravenous methylprednisolone twice daily and oral vancomycin for 10 days, which resulted in symptomatic improvement and the absence of any pathogens from her repeat stool microbiological investigations. Prior to discharge, her renal ultrasound was normal and CA-125 was 21.8 U/mL. The patient was discharged from hospital with 12 mg oral prednisolone and 0.2 g hydroxychloroquine. At follow up one month later, there was no vomiting nor diarrhea.

### **DISCUSSION**

CA125 is a biomarker for gynaecological malignancy. Clinically, CA125 can be elevated by various benign diseases. Elevated CA-125 Levels in SLE patients are attributed to mesothelial cell activation. In SLE patients, elevated serum CA125 Levels are independently associated with serositis<sup>2, 3</sup>. Pleural and pericardial effusions are not uncommon among SLE patients. However, massive ascites are rare in SLE patients without any other complications<sup>4</sup>. Ascites in SLE are attributed to nephrotic syndrome, constrictive pericarditis, lupus peritonitis, protein-losing enteropathy or Budd-Chiari syndrome. A rapid onset of massive ascites can be an initial manifestation of SLE<sup>5</sup>. Our patient presented with painless massive ascites co-existing with low complement and hypoproteinemia. However, she did not show any overt proteinuria, and heart ultrasound as well as hepatic hilum ultrasound were normal. Therefore, lupus peritonitis, nephrotic syndrome, constrictive pericarditis and Budd-Chiari syndrome were ruled out. We postulated that hypoproteinemia was due to protein-losing enteropathy (PLE), resulting in intestinal damage caused by SLE (diarrhea, bowel wall edema and mesenteric vasculitis), consistent with previous studies<sup>6, 7</sup>. However, 99mTc-HAS (99m-labeled human serum albumin) is required for definite diagnosis<sup>8</sup>, which is not available at our hospital. Lupus cystitis is a rare complication of SLE that generally presents with lower urinary tract symptoms and gastrointestinal symptoms, such as vomiting, nausea, and abdominal pain<sup>9, 10</sup>. Ultrasonographic examination of the patient showed bilateral hydronephrosis and hydroureter in addition to bladder wall thickening, which conforms to manifestations of lupus cystitis. Yuan *et al*<sup>11</sup> reported that lupus mesenteric vasculitis and lupus cystitis concurrently occurred in 22.7% of patients, thus, lupus cystitis should be suspected in SLE patients, especially those with lower urinary tract and gastrointestinal symptoms.

We summarized the clinical features of previous 20 cases of Tjalma syndrome and current cases (Table 1). All patients were female, and their mean age was  $36.5 \pm 10.7$  (mean $\pm$ SD) years. A decrease in serum C3 and C4 Levels was reported in all Tjalma syndrome patients, which was attributed to complement consumption caused by complement system activation<sup>12</sup>. The patient was clinically diagnosed with SLE with

elevated CA-125, but there were no benign or malignant tumors. A review of previous studies revealed ascites and pleural effusions in all cases, but only 10 patients presented with pericardial effusions. Although there were no pleural effusions, just as pericardial effusions were not found in some previous cases, the clinical features of this case fit the Tjalma syndrome, which can be a specific finding. Tjalma syndrome can present with massive ascites alone without pleural or pericardial effusions, which requires further clinical attention. Generally, the Tjalma syndrome has good prognostic outcomes after administration of methylprednisolone and immunosuppressants, with resolution of ascites and pleural effusions, and normalization of CA-125.

### **CONCLUSION**

In conclusion, massive ascites with increased CA125 do not always indicate the presence of malignancy, especially in patients with SLE. Although rare, the Tjalma syndrome has been increasingly reported in recent years, therefore, there is a need for increased awareness of this condition.



4%

SIMILARITY INDEX

PRIMARY SOURCES

1	<a href="http://www.wjgnet.com">www.wjgnet.com</a> Internet	24 words — 2%
2	Fei Gao, YongMei Xu, GuoWang Yang. "Pseudo-pseudo Meigs' syndrome presenting with a combination of polyserositis, elevated serum CA 125 in systemic lupus erythematosus", Medicine, 2019 Crossref	21 words — 1%
3	<a href="http://Www.Wjgnet.Com">Www.Wjgnet.Com</a> Internet	13 words — 1%

EXCLUDE QUOTES ON  
EXCLUDE BIBLIOGRAPHY ON

EXCLUDE SOURCES < 12 WORDS  
EXCLUDE MATCHES < 12 WORDS