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

Wang JD *et al*. Tjalma syndrome

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

**Jun-Di Wang, Xian-Feng Zhang, Jiao Huang,** Department of Rheumatic Disease, Affiliated Hangzhou First People’s Hospital, Zhejiang University School of Medicine, Hangzhou 310000, Zhejiang Province, China

**Yan-Fei Yang,** Department of Respiratory Disease, Hangzhou Hospital of Traditional Chinese Medicine, Hangzhou 310000, Zhejiang Province, China

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**Corresponding author: Jiao Huang, MD, Associate Chief Physician,** Department of Rheumatic Disease, Affiliated Hangzhou First People’s Hospital, Zhejiang University School of Medicine, No. 261 Huansha Road, Hangzhou 310000, Zhejiang Province, China. huangjiao2001@163.com

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**Abstract**

BACKGROUND

Ascites, pleural effusion and raised CA-125 in the absence of malignancy in systemic lupus erythematosus is known as Tjalma syndrome.

CASE SUMMARY

We report a special case of a systemic lupus erythematosus patient presenting with Tjalma syndrome. 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; Pseudo–pseudo Meigs’ syndrome; Systemic lupus erythematosus; Ascites, CA-125, Case report

Wang JD, Yang YF, Zhang XF, Huang J. Systemic lupus erythematosus presenting with progressive massive ascites and CA-125 elevation indicating Tjalma syndrome? A case report. *World J Clin Cases* 2022; In press

**Core Tip:** We report a special case of a systemic lupus erythematosus patient presenting with pseudo–pseudo Meigs’ syndrome. 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 (SLE) is a chronic, autoimmune disease with multiple systemic disorders. Tjalma syndrome, also known as pseudo–pseudo Meigs’ syndrome, 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[1]. Massive ascites are rare in SLE patients without any other complications. Herein we report a special case of an SLE patient presenting with Tjalma syndrome. She presented with ascites and elevated CA-125 but no pleural effusions.

**CASE PRESENTATION**

***Chief complaints***

A 23-year-old woman presented 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 d ago where physical examination revealed a distended abdominal wall, while abdominal computed tomography scan revealed massive ascites (Figure 1A). Laboratory examinations at the outside hospital showed markedly elevated CA-125 at 1685 U/mL (0-35 U/mL). Ascitic fluid analyses revealed negative results from Rivalta tests. After diuresis treatment for 7 d, the amount of ascites in the patient was gradually reduced. However, there were no changes in nausea or vomiting.

***History of past illness***

The patient had a history of immune thrombocytopenia for 2 years and was administered with a long-term maintenance dose of 5 mg/d prednisone.

***Personal and family history***

The patient denied any family history.

***Physical examination***

Patient temperature and blood pressure were 37.2 ºC 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***

The following is the patient’s laboratory examination results: White blood cell count, 6.8 × 109/L; neutrophil%, 77.9%; hemoglobin, 100  g/L; platelet, 130 ×  109/L; total protein, 58 g/L; albumin, 31.6  g/L; d-dimer, 1910 μg/L; and ferritin, 37.7 μg/L. The 24-h urine protein quantitate was 74  mg/24  h. Antinuclear antibody 1:100 (+), Anti-Sjogren’s syndrome A antigen antibody (+), 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 ×  106/L; T-cell lymphocytes (CD3), 226 ×  106/L; B-cell lymphocytes (CD19), 265.9 ×  106/L; T-helper lymphocytes (CD4), 54.4 ×  106/L; natural killer cells (CD16/56), 5.6 ×  106/L; and CD4/CD8, 0.71. In addition, the tumor marker (CA-125) was 439.9 U/mL, whereas other tumor markers, including AFP, CEA, NSE, CA153, CA199 and β-HCG were normal. Moreover, erythrocyte sedimentation rate, C-reactive protein, ANCA, index of autoimmune liver diseases, immunoglobulin G4, hepatitis B surface antigen and HIV were all found to be within normal ranges. T-SPOT showed negative results.

***Imaging examinations***

Small bowel enhanced computed tomography 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 1B). Enhanced magnetic resonance imaging 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 and lupus cystitis.

**TREATMENT**

The patient was treated with 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 d 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 computed tomography 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 d, 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 the hospital with 12 mg oral prednisolone and 0.2 g hydroxychloroquine. At follow up 1 mo later, there was no vomiting or diarrhea.

**DISCUSSION**

CA-125 is a biomarker for gynecological malignancy. Clinically, CA-125 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 CA-125 levels are independently associated with serositis[2,3]. Pleural and pericardial effusions are common among SLE patients. However, massive ascites are rare in SLE patients without any other complications[4]. 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[5].

Our patient presented with painless massive ascites coexisting 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, resulting in intestinal damage caused by SLE (diarrhea, bowel wall edema and mesenteric vasculitis), consistent with previous studies[6,7]. However, 99m-labeled human serum albumin is required for definite diagnosis[8], 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[9,10]. 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*[11] 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 ± 10.7 (mean ± 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[12]. 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, 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 CA-125 do not always indicate the presence of malignancy, especially in patients with SLE. Although rare, Tjalma syndrome has been increasingly reported in recent years. Therefore, there is a need for increased awareness of this condition.

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**Footnotes**

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**Figure Legends**



**Figure 1 Computed tomography scan.** A: Abdominal computed tomography scan revealed massive ascites (orange arrow); B: Small bowel enhanced computed tomography revealed that the number of mesenteric vessels was increased. Mesenteric vessels were engorged and exhibited a “comb sign” (orange arrow) appearance.

**Table 1 Reported cases of pseudo-pseudo Meigs syndrome**

|  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Year** | **Ref.** | **Gender** | **Age** | **Naive SLE** | **CA125**1 | **Ascites** | **Pleural effusion** | **Pericardial effusion** | **Nausea/vomiting** | **Dyspnea** | **Hypoproteinemia** | **Proteinuria** | **ANA** | **dsDNA** | **SSA** | **Low complement**  | **Leukopenia** | **Anemia** | **Thrombocytopenia** | **APS** | **Treatment** | **Outcome** |
| 2005 | Tjalma[13] | Female | 38 | Yes | 887 | + | + |  |  | + | + |  |  |  |  |  |  | + | + | + | MP + AZA | Remission  |
| 2005 | Schmitt *et al*[1] | Female | 33 | Yes | 2287 | + | + |  |  | + | + | + | + | + |  | + | + | + | + | + | MP + MMF + HCQ | Remission  |
| 2008 | Ural *et al*[14] | Female | 38 | Yes | 1229 | + | + |  |  | + |  |  | + | + | + | + |  |  |  |  | MP + HCQ | Remission  |
| 2011 | Besand Soy[15] | Female | 47 | Yes | 233 | + | + | + | + | + |  |  | + | + |  | + | + | + |  |  | MP + HCQ | Remission  |
| 2012 | Dalvi *et al*[3] | Female | 56 | No | 70.1 | + | + |  | + |  |  |  | + | + |  | + | + |  |  |  | MP + MMF | Remission  |
| 2013 | Bes *et al*[16] | Female | 42 | Yes | 91.3 | + | + | + | + | + | + | + | + | + |  | + |  |  | + |  | MP + CYC + AZA | Remission  |
| 2013 | Lee *et al*[17] | Female | 29 | Yes | 345 | + | + | + | + | + | + | + | + | + |  | + | + | + | + | + | MP + HCQ | Remission  |
| 2013 | Lee *et al*[17] | Female | 54 | No | 344.9 | + | + | + |  |  | + | + | + | + |  | + | + |  | + |  | MP + MMF | Remission  |
| 2016 | Cheah *et al*[7] | Female | 34 | No | 1613.8 | + | + |  |  |  | + |  | + |  |  | + |  | + |  |  | MP + HCQ | Remission  |
| 2016 | McVorran *et al*[18] | Female | 40 | Yes | 307 | + | + | + |  | + |  |  | + | + |  | + |  |  |  |  | MP | Remission  |
| 2019 | Torres Jiménez *et al*[19] | Female | 14 | Yes | 59 | + | + | + |  | + | + |  | + | + |  | + |  | + | + |  | MP + CYC + MMF + RTX | Remission  |
| 2018 | Zampeli *et al*[20] | Female | 40 | No | 85 | + | + | + |  | + | + |  | + | + | + | + | + | + |  |  | MP + CYC + MMF | Remission  |
| 2019 | Awad *et al*[21] | Female | 43 | No | 80 | + | + |  |  |  | + | + | + | + |  | + |  | + |  |  | MP + MMF + HCQ | Remission  |
| 2019 | Tansir *et al*[22] | Female | 22 | Yes | 2025 | + | + | + |  |  | + | + | + | + |  | + |  | + | + | + | MP + CYC + HCQ + AZA | Remission  |
| 2019 | Li *et al*[23] | Female | 24 | No | 949 | + | + | + |  | + | + |  | + |  |  | + | + |  |  |  | MP + MMF | Remission  |
| 2019 | Ahmed *et al*[24] | Female | 44 | Yes | 227 | + | + |  | + |  | + |  | + | + | + | + |  |  |  |  | MP + AZA | Remission  |
| 2019 | Gao *et al*[6] | Female | 44 | Yes | 360.8 | + | + |  |  | + | + | + | + | + | + | + | + | + |  |  | MP + HCQ + LEF | Remission  |
| 2021 | Quintero-Muñoz *et al*[25] | Female | 33 | No | 187 | + | + |  | + | + | + | + | + | + | + | + | + |  |  | + | MP + MMF + HCQ + CYC | Death |
| 2021 | Meena *et al*[26] | Female | 23 | No | 230.5 | + | + |  |  | + | + |  | + | + |  | + |  | + |  | + | MP + HCQ + AZA | Remission  |
| 2022 | Karadeniz *et al*[27] | Female | 33 | No | 476 | + | + | + | + | + | + | + | + | + |  | + |  |  |  |  | MP + MMF + HCQ | Remission  |
| 2022 | Current case | Female | 23 | Yes | 1685 | + |  |  | + |  | + |  | + |  | + | + |  |  | + |  | MP + HCQ | Remission  |

1U/mL.

ANA: Antinuclear antibodies; dsDNA: Anti-double-stranded DNA antibodies; SSA: Anti-Sjögren's syndrome-related antigen A; APS: Antiphospholipid syndrome; MP: Methylprednisolone; AZA: Azathioprine; MMF: Mycophenolate mofetil; HCQ: Hydroxychloroquine; CYC: Cyclophosphamide; RTX: Rituximab; LEF: Leflunomide.