Weight Loss, Ascites, and Diarrhea as the Presenting Symptoms for Pseudo-Pseudo Meigs Syndrome

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Background

- **Pseudo-pseudo Meigs syndrome (PPMS) is a rare presentation of systemic lupus erythematous (SLE) characterized by ascites, pleural effusion, and elevated CA-125 level.**

Case

A 30-year-old woman with SLE and history of small bowel obstruction presented with diarrhea, weight loss, anasarca, and alopecia.

Physical exam: she was tachycardic and hypertensive. Her exam revealed profound anasarca with ascites and bilateral pleural effusions.

Cr=5.65 mg/dL (baseline 0.7 mg/dl)	C3=49 mg/dL C4=19 mg/dL
Spot protein/creatinine =3.9	CA-125=260.8 U/mL
ferritin=554.6 ng/ml	+ANA
ESR=145 mm/Hr	dsDNA=356 U/ml
CRP=6 mm/Hr	+pANCA/MPO

Workup

- Ovarian malignancy was ruled out by **MRI and CT abdomen/pelvis**.
- Inflammatory bowel disease was ruled out by **EGD and colonoscopy**.
- **Renal ultrasound** showed bilateral hydronephrosis.
- **Transvaginal ultrasound** was normal.
- **Paracentesis**: SAAG 1.3.
- **Thoracentesis**: total serum protein 4.2 g/dl | pleural fluid LDH 67 U/L.
- **Kidney biopsy** demonstrated class IV / V lupus nephritis.

Treatment

She was treated with a 3-day course of pulse dose methylprednisolone followed by prednisone, hydroxychloroquine, and furosemide infusion. Her symptoms mostly resolved and CA-125 trended down to 27 U/mL.

Discussion

- This case illustrates the challenges of evaluating abdominal symptoms and ascites in patients with SLE and the importance of a kidney biopsy in the evaluation of nephrotic range proteinuria.
- **Due to infrequency and concern for malignancy due to elevated CA-125 level, patients with this syndrome undergo extensive workup.** Imaging may exclude mesenteric vasculitis, an uncommon feature of SLE which can be seen as targetoid lesions of the bowel on CT scan.
- CA-125 has been shown to be elevated due to inflammatory markers enhancing its expression in peritoneal mesothelial cells. **Prior publications have shown the correlation of CA-125 with extent of serositis in PPMS.**
- Our patient also has many other clinical features reported in PPMS patients including elevated ferritin, alopecia, and positive pANCA. CT scan of abdomen and pelvis is essential in the workup of this syndrome to rule out ovarian malignancy.
- Steroids are the treatment for PPMS.