Intestinal (pseudo-)obstruction and hydronephrosis – consider lupus

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A 57-year-old woman diagnosed with systemic lupus erythematosus (SLE) in 2005 was admitted to the rheumatology department due to abdominal pain, nausea, and vomiting.

The initial diagnosis of SLE in 2005 was based on arthralgia, Raynaud's phenomenon, mucosal erosion, and pleural effusion. Typical laboratory findings included elevated anti-dsDNA antibody levels and decreased C3 and C4 complement levels. Throughout the course of the disease, the patient primarily exhibited musculoskeletal symptoms and mild serositis. Since diagnosis, treatment with variable doses of prednisone (5-20mg/d) was administered. Attempts to introduce immunomodulatory drugs were unsuccessful due to adverse events; hydroxychloroquine treatment was discontinued due to blurred vision, and methotrexate was halted due to gastrointestinal symptoms.

Upon admission with gastroenterological symptoms, the patient was solely on prednisone 5mg/d. Despite symptomatic treatment with spasmolytic and antiemetic medications, the patient's condition deteriorated – manifesting as increased abdominal pain, cessation of intestinal peristalsis, and oliguria. Laboratory tests revealed elevated CRP (19 mg/l, norm <5 mg/l), lymphopenia (0.58 x 10^3/ul, norm <1.3 x 10^3/ul), decreased complement components (C3 – 69 mg/dl, norm 90-180 mg/dl; C4 8 mg/dl, norm 10-40 mg/dl), and a high level of anti-dsDNA antibodies (>666.9 IU/dl). Abdominal X-ray showed no features of intestinal obstruction (like dilated bowel loops, or the presence of fluid levels) or perforation (like free gas under the diaphragm). Abdominal ultrasound showed segmental intestinal wall thickening (Figure 1A) and bilateral hydronephrosis (Figure 1B). A subsequent CT scan demonstrated thickening and post-contrast enhancement of the walls of the stomach, duodenum, and small intestine, with obliteration of the surrounding fat tissue (Figure 1C), and a slight enlargement and enhancement of the common bile duct (Figure 1D). After exclusion of mechanical causes of intestinal obstruction (no distinct transition point where bowel caliber changes from normal to abnormal, signs of twisting of the mesentery, or bowel strangulation in imaging tests), the patient was diagnosed with intestinal pseudo-obstruction (IPO) in the course of SLE. The patient received 1 mg/kg/day IV methylprednisolone for 3 days, followed by prednisone 0.5mg/kg/day PO, leading to symptom resolution.

IPO is a rare complication of SLE, affecting up to 2% of SLE patients[1]. It is more prevalent in young (<40 years) females, often presenting early in the disease course and in patients with affected hematological domain, polyserositis, and hypocomplementemia[1,2]. The pathogenesis of IPO is not fully understood, but it is assumed to be related to systemic smooth muscle

involvement. Symptoms of intestinal obstruction dominate in the clinical picture, accompanied by hydronephrosis, and occasionally megacholedochus[1]. First-line treatment is high-dose glucocorticoids, with an 80% positive response rate[1,2]. In cases of glucocorticoid ineffectiveness, immunomodulatory drugs like cyclophosphamide, hydroxychloroquine, azathioprine, cyclosporine, or tacrolimus can be considered. Despite its rarity, awareness of IPO is crucial, as it is misdiagnosed in approximately 40% of SLE patients and up to 80% if IPO is the initial manifestation of SLE[1,2].

Tables and Figures

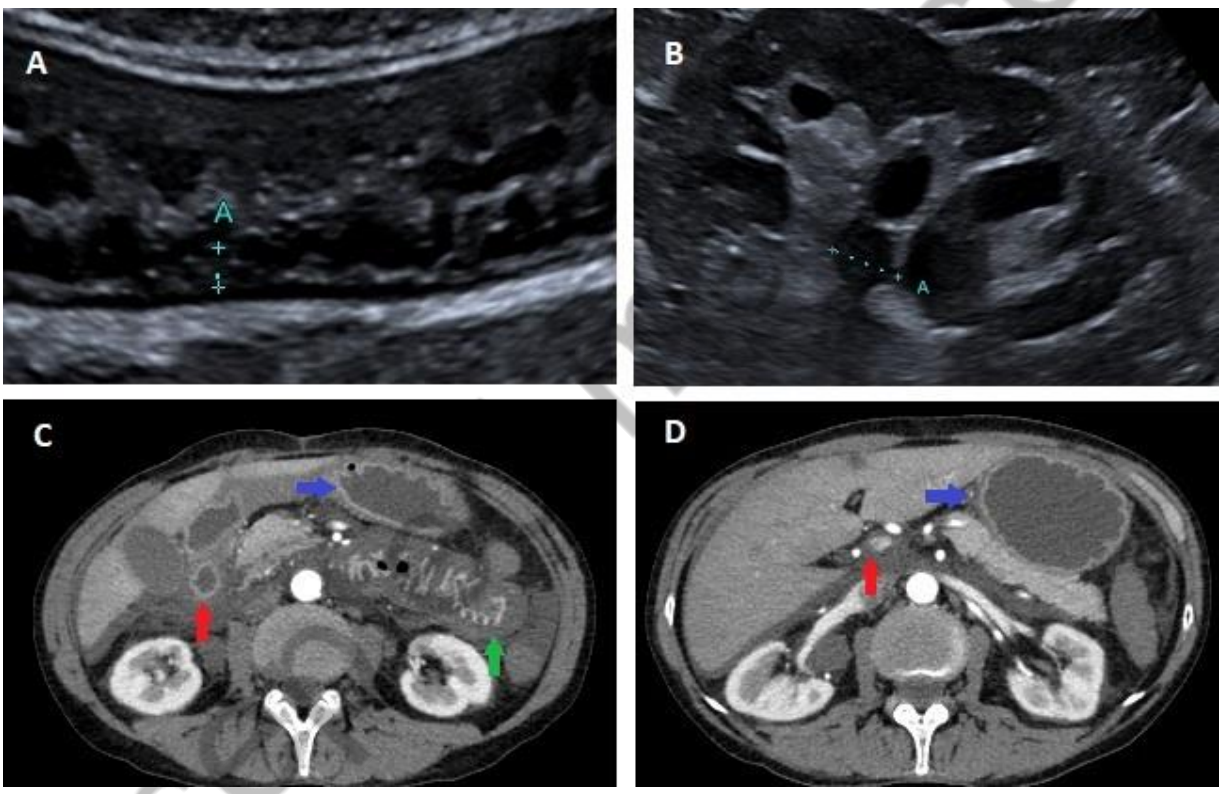

Figure 1. Intestinal pseudo-obstruction presentation. A) Segmental intestinal wall thickening in abdominal ultrasound. B) Hydronephrosis in abdominal ultrasound. C) Thickening and post-contrast enhancement of the walls of the stomach (blue arrow), duodenum (red arrow), and small intestine (green arrow), with obliteration of the surrounding fat tissue in abdominal computed tomography. D) Slight enlargement and enhancement of the common bile duct (red arrow) and thickening and post-contrast enhancement of the walls of the stomach (blue arrow) in abdominal computed tomography.

References

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