CASE REPORT

Intestinal pseudo-obstruction and ureterohydronephrosis as the presenting manifestations of relapse in a lupus patient

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Intestinal pseudo-obstruction (IPO) is a rare complication of systemic lupus erythematosus (SLE). We present a 32-year old female with SLE for seven years. She was admitted with profound fatigue, frequent vomiting, colicky abdominal pain, diarrhoea and intermittent dysuria for the past 12 months. Imaging studies revealed dilated small and large bowel loops with thickened intestinal wall and multiple fluid levels. Urinary tract involvement was also demonstrated. The patient responded well to immunosuppressive treatment. IPO in the context of SLE has been described only in anecdotal case reports. Half of the cases developed this complication during the course of lupus as in the present case. Concomitant ureterohydronephrosis was present in approximately two-thirds of the cases. Early recognition of the syndrome is necessary for the institution of the appropriate medical treatment and prevention of inappropriate surgical intervention. *Lupus (2004) 13, 954–956.

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Intestinal pseudo-obstruction (IPO) is a recently recognized manifestation of systemic lupus erythematosus (SLE).1 IPO is commonly encountered with urologic complications designated as ureterohydronephrosis and/or interstitial cystitis.2 A case of IPO in a patient with underlying SLE is reported and the clinical, laboratory and pathogenetic features are discussed.

A 32-year old Asian woman was first seen seven years ago with malar rash, fever, arthralgias, leucopenia and thrombocytopenia. SLE was diagnosed and she started on corticosteroids and hydroxychloroquine with remission of the symptoms. She never experienced arterial or venous clotting and had never been pregnant. She remained stable for six years but was then readmitted with profound fatigue, frequent vomiting (10 episodes per day) intractable to anti-emetics, colicky abdominal pain and diarrhoea for the past 12 months. She also complained of intermittent dysuria with negative bacterial and atypical bacterial urine cultures. She had several hospitalizations during the last 12 months where she received anti-emetics and omeprazole without remission of the symptoms. Her medication on admission were methylprednisolone 8 mg daily, hydroxychloroquine 200 mg daily, diclofenac 50 mg thrice daily and ofloxacin 200 mg twice daily. Her symptoms worsened and she reported a loss of weight of 20 kg over the last few months before admission to our department. On examination she looked ill, and was underweight (BMI 17.3 kg/m²). She was fully oriented and afebrile. Her blood pressure and pulse rate were normal. She had alopecia and facial swelling. Abdominal distention and tenderness with sluggish bowel sounds were present. She was found to have lymphopenia, 0.36 × 10⁹/L and proteinuria 0.97 g/24 h. Urinalysis revealed dysmorphic red blood cells and leukocytes and a sterile culture. ESR was 67 mm/h and CRP was less than 5 mg/L. Antinuclear antibodies were positive in high titer > 1/640, anti-double-stranded DNA (anti-dsDNA) antibodies were positive 37 U/mL (normal values < 17 U/mL), anti-RNP antibodies were positive, β₂ glycoprotein I IgM were 200 U/mL (normal values < 100 U/mL), C3, C4 were low, 47.5 mg/dL (normal 55–120 mg/dL) and 11 mg/dL (20–50 mg/dL), respectively. Anticardiolipin antibodies and lupus anticoagulant were negative. Colonoscopy was normal. Computed Tomography (CT) scan of the abdomen disclosed thickening of the intestinal wall and dilated loops of the small bowel and colon. Hydroureter and hydronephrosis were also revealed (Figures 1 and 2). Cystoscopy examination was normal and bladder biopsies disclosed no interstitial cystitis. A relapse of SLE was diagnosed with renal involvement, coexisting with IPO and ureterohydronephrosis. Hydroxychloroquine, diclofenac...
and ofloxacin were stopped and treatment with prednisolone 1 mg/kg/daily and intermittent pulses of cyclophosphamide 20 mg/kg/monthly were initiated. Three weeks following onset of treatment, the patient’s symptoms and signs settled and she was able to eat without vomiting. Corticosteroids were tapered gradually over the subsequent five months to 8 and 12 mg on alternate days. Five months following the onset of treatment, she has gained 10 kg in weight. Complement values reverted towards normal, the antinuclear antibody titer fell to 1/640 and anti-dsDNA fell to 9 U/mL. Imaging findings of the bowel reverted to normal and urinary tract dilatation improved (Figure 3).

Gastrointestinal manifestations are common in SLE. IPO is an uncommon complication of lupus and is defined as the presence of clinical features of intestinal obstruction without an identifiable organic cause. Vasculitis leading to chronic ischemia of the bowel has been considered as a possible pathogenetic mechanism. Another cause that has been postulated is an intrinsic muscle dysmotility affecting the muscularis propria. IPO is associated with the urological complication of ureterohydronephrosis in 63.6% of the cases. The apparently high association between these two conditions suggests a possible common smooth muscle dysmotility due to primary myopathy or neurogenic pathology secondary to either immune-complex mediated vasculitis or common circulating autoantibodies against smooth muscle.

The diagnosis of this entity is based on imaging findings consistent with the presence of dilated small and/or large bowel loops with thickened intestinal wall and multiple fluid levels. Gastrointestinal manometric studies may help in diagnosis. Concomitant ureterohydronephrosis may be present and histological examination of the urinary bladder may reveal interstitial cystitis in 31.8% of cases. Our patient had urinary tract involvement in the form of hydronephrosis, dilatation of the ureter and thickening of the ureter wall.

Most of the information on this newly recognized clinical entity is arising from anecdotal case reports. Two studies in the literature comprise description of new cases in combination with review of the literature. From 18 and 21 patients with IPO and underlying lupus, 50 and 41% had IPO as the initial presentation and the remaining 50 and 59% of the cases, respectively, developed IPO as a complication of underlying SLE as in the present case. In the latter group, the mean duration of lupus prior to the diagnosis of IPO was 8 ± 10 years (range 1–36 years). Our patient developed IPO six years following the diagnosis of SLE.
The manifestation of IPO in our patient was associated with an active lupus serology, a feature present in the majority of cases reported in the literature.\(^1\) Active lupus serology, low C3 and C4 levels, findings in the urine sediment consistent with glomerulonephritis and response to immunosuppressive treatment suggest an immune-complex mediated mechanism of the syndrome. The patient was positive for $\beta_2$-glycoprotein I antibodies but negative for anticardiolipin antibodies and lupus anti-coagulant and she never experienced an arterial or venous clotting. Hitherto, antiphospholipid antibody syndrome was not documented and it was probably not involved in the current manifestation of lupus.

High dose steroids are the treatment of choice in patients with IPO and urinary tract involvement leading to clinical remission and disappearance of imaging findings.\(^5,6\) In the current case, symptoms from IPO and urinary tract involvement were dramatically settled. The thickening of the intestinal wall and ureter and air fluid levels were all reverted towards normal after treatment and ureterohydronephrosis was ameliorated. The high reversibility of symptoms and findings under medical treatment is demonstrated in current case and is emphasized in literature. Two cases in the literature were treated with pulses of cyclophosphamide as a maintenance agent.\(^5\) Because of coexisting renal involvement in the current case, the addition of cyclophosphamide to steroids was deemed necessary.

IPO is a recently recognized rare complication of SLE. Hence, the diagnosis is usually delayed and the clinical condition of the patients is poor. It is noteworthy that 10 of 18 patients reported in the literature underwent surgical intervention because of symptoms of intestinal obstruction.\(^1\)

In conclusion, IPO may occur concomitantly with urinary tract involvement and is a severe manifestation of SLE with high morbidity. A high level of clinical awareness is essential for preventing inappropriate surgical intervention. Early recognition of the syndrome is necessary for the institution of the appropriate medical treatment.

References