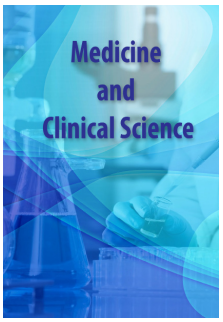
Medicine & Clinical Science

Case Report



Abdominal distension and vomiting in a lupus patient with a desire for pregnancy: think beyond the baby!

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Abstract

Intestinal pseudo-obstruction (IPO) is a rare enteric motility disorder with clinical features of intestinal obstruction in the absence of mechanical blockage. IPO usually presents as recurrent, self-limiting episodes of abdominal distension, postprandial pain, nausea and vomiting. Over time, symptoms can persist, leading to gut stasis with bacterial overgrowth, malabsorption and malnutrition. We present a case of IPO related to systemic lupus erythematosus, treated with intravenous pulses of high-dose methylprednisolone. Nutritional testing should be provided as insidious progression to intestinal failure may go undetected at an early stage, especially in situations with increased nutritional requirements, like pregnancy.

Introduction

Intestinal pseudo-obstruction (IPO) is a syndrome with clinical features of intestinal obstruction in the absence of a mechanical cause [1]. This rare but severe enteric motility disorder is probably under recognized and underreported with an estimated prevalence of <1/100,000 [2]. The pathophysiology of IPO is poorly understood. It can be classified according to etiology (primary, secondary or idiopathic) or histopathology - differentiating between myopathies, neuropathies and mesenchymopathies based upon the predominant involvement of smooth muscle cells, enteric neurons or the interstitial cells of Cajal respectively. Overlap between these entities is common [3].

Most cases initially present with recurrent, subacute, self-limiting episodes. These episodes are characterized by marked abdominal distension and/or severe postprandial pain accompanied by nausea and vomiting. Over time, symptoms no longer subside, their persistent character leads to gut stasis with bacterial overgrowth, malabsorption and malnutrition [1].

Diagnosis is often delayed due to self-limiting nature of early disease course and can be preceded by unnecessary surgical interventions. Clinical recognition is key, further supported by laboratory, radiology, manometry and histopathology findings. To date, there are no established diagnostic criteria for IPO [4]. Treatment should target the underlying pathology, together with symptom management and nutritional support. The disease course usually leads to intestinal failure, requiring parenteral nutrition or even intestinal transplantation [1,5].

Systemic lupus erythematosus (SLE) is one of the possible secondary causes of IPO. SLE is an autoimmune disease potentially affecting all organ systems. Involvement of the skin, joints, kidneys and blood vessels is well recognized, while gastrointestinal manifestations often attract less attention [6]. We report a patient with IPO related to SLE who was effectively treated with high-dose corticosteroids.

Case Report

A 39-year-old woman of Arab origin consulted our emergency department with abdominal pain, nausea, vomiting and absence of flatus for three days. Six weeks beforehand, she underwent fertility treatment, followed by miscarriage in the first trimester of pregnancy. Four weeks after the miscarriage, she suffered an unexplained tonic-clonic seizure. Her medical record revealed autoimmune thyroiditis and SLE (diagnosed in the United Arab Emirates) with association of an antiphospholipid syndrome (APS). There was no history of abdominal nor pelvic surgery. She was currently taking prednisolone 5 mg daily and thyroid hormone therapy.

On admission, her vital signs were normal. Her abdomen was markedly distended with diffuse tenderness and scarce bowel sounds. Biology showed a C-reactive protein of 66.7 mg/L, erythrocyte sedimentation rate of 102 mm/h and leucocyte count of 6.1 x 109/L with neutrophil predominance. Liver function tests, lipase, creatinine kinase, lactate dehydrogenase and thyroid function tests were all normal.

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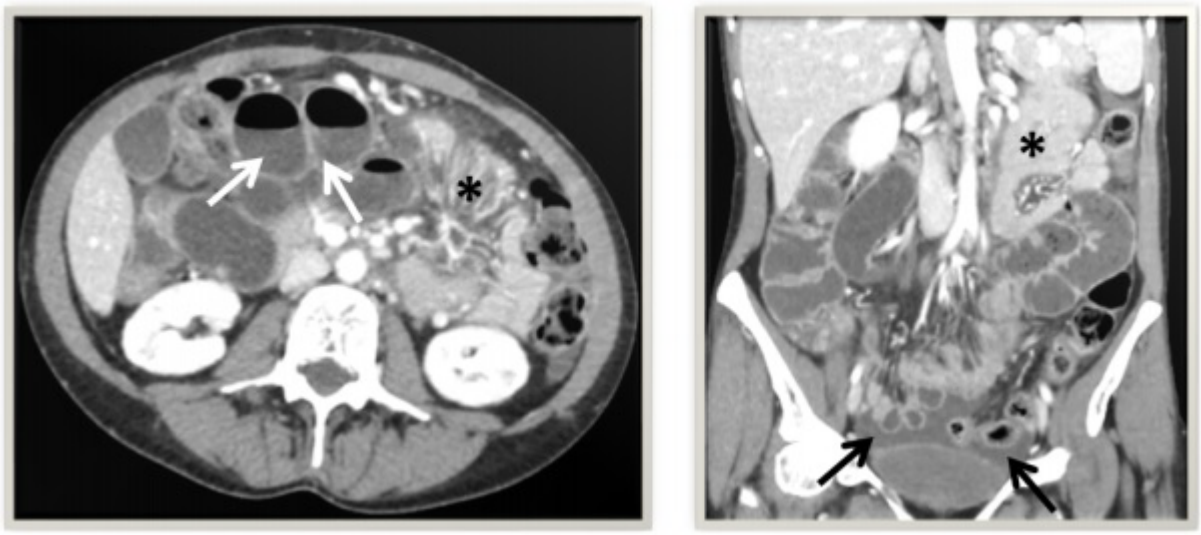

Figure 1. Computed tomography.
Abdominal CT scan revealing a collapse of the proximal and distal small intestine (asterisk) with multiple changes in caliber, air-fluid levels (white arrow) and small amounts of ascites (black arrow). This is compatible with intestinal pseudo-obstruction.

Abdominal CT scan revealed a collapse of the proximal and distal small intestine with multiple caliber changes, air-fluid levels and a small amount of ascites (Figure 1). There was no sign of intestinal ischemia as bowel wall enhancement was normal.

Initial management consisted of intravenous hydration and nasogastric decompression. Intestinal pseudo-obstruction due to active SLE was considered the most likely diagnosis. Intravenous pulses of high-dose methylprednisolone (500 mg) were initiated for three consecutive days, tapered to 32 mg orally. Thromboprophylaxis with low molecular weight heparin was given for five days. Clinical evolution was favorable. She was discharged six days after admission. Follow-up with colonoscopy was planned. Unfortunately, the patient was lost to follow-up.

Discussion

We present a patient with IPO as a gastrointestinal complication of SLE. The presence of a well-defined diagnosis of lupus was essential in making a timely diagnosis. Furthermore, the recent history of new-onset epilepsy and miscarriage were indicating active lupus, despite low-dose prednisolone maintenance therapy.

Mechanical obstruction was deemed unlikely due to presence of caliber changes at multiple levels on abdominal imaging. Lupus mesenteric vasculitis was another possible SLE-related complication, able to mimic the image of pseudo-obstruction, especially in the context of APS. However, the normal bowel wall enhancement and three-day history of mild abdominal pain made this diagnosis less likely [4,7].

Hypothyroidism was also considered given the history of autoimmune thyroiditis. The thyroid function was correctly substituted. In light of this deduction and in absence of established diagnostic criteria, we presumed the diagnosis of SLE-IPO.

IPO is a rare manifestation of SLE with an estimated prevalence of 2% [8]. The exact pathogenesis is unknown. No 'SLE-IPO specific auto-antibodies' are known to date, although a predominance for anti-SSA antibodies is suggested. In half of the cases, IPO is the initial manifestation of SLE, while the other half is associated with poorly controlled systemic disease [9]. Mortality rate in SLE-IPO is high (7%) as a consequence of bowel perforation and sepsis, if not treated promptly with immunosuppressive agents. Relapse is often related to noncompliance. Nonetheless, progression to chronic IPO (CIPO) is possible, with development of bowel wall atrophy and fibrosis, leading to intestinal failure [4]. Nutritional assessment and -support is crucial [1].

Little is known about SLE-IPO in the context of pregnancy or fertility treatment. Active SLE is associated with pregnancy loss and optimal control prior to conception should therefore be pursued [10]. Maternal micronutrient deficiencies and malabsorption are also risk factors for pregnancy loss.

Hypothetically, this could be a vicious circle for SLE-IPO patients with the desire for pregnancy, as pregnancy may provoke IPO flares in SLE patients, gradually leading to malabsorption, in turn predisposing to pregnancy failure. New pregnancy attempts could provoke the progression to CIPO,resulting in increased risk of miscarriage. This hypothesis is depicted in Figure 2. It is unclear whether our patient was already suffering some form of malabsorption, as she was lost to follow-up.

In conclusion, we presented a patient with intestinal pseudo-obstruction associated with systemic lupus erythematosus. IPO is a rare but potentially lethal complication of SLE, especially if misdiagnosed and adequate treatment is postponed. Initial therapy consists of high-dose corticosteroids, with a good response in the majority of cases. Insidious disease progression may gradually lead towards intestinal failure, making regular nutritional screening and support essential, especially in situations with increased risk of malabsorption, such as pregnancy.

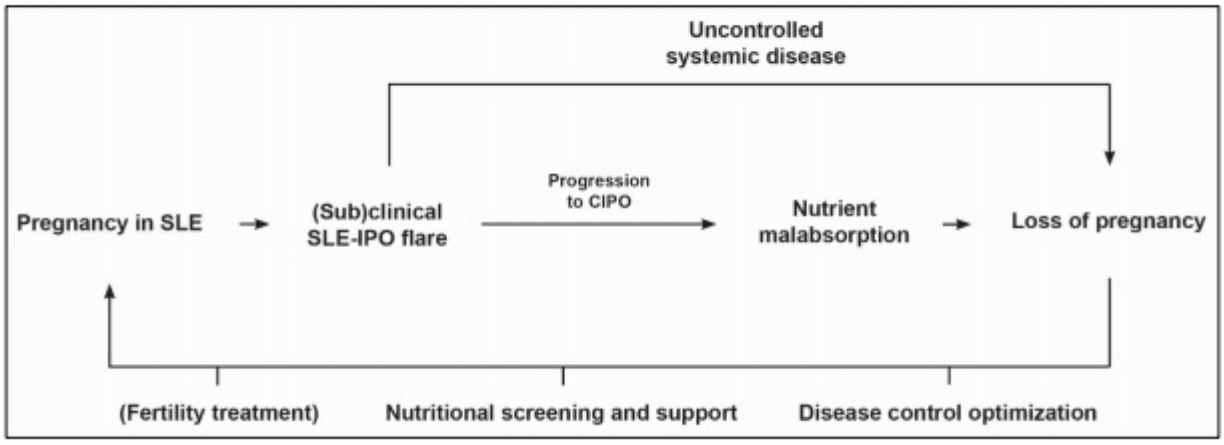

Figure 2. Hypothesis of the vicious circle of pregnancy loss in SLE-IPO.
Pregnancy could provoke a flare of SLE-IPO, which may go unnoticed especially in gravid patients. Active SLE may predispose to miscarriage. Additionally, subclinical IPO flares may progress towards irreversible CIPO, where nutrient malabsorption will further increase the risk of pregnancy loss. Optimal disease control together with vigorous nutritional screening and support should therefore be provided prior to fertility treatment in SLE patients, in order to ensure safe pregnancy and avoid progression towards CIPO.

Competing interests

The authors declare no conflict of interest.

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Contributions from authors

GR, TVN and JdF equally contributed to the draft. MS contributed to the discussion and final revision of the paper. HR contributed to the revision of the draft and supervised the writing process.

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Prior presentation

The case was presented by Jeroen de Filette as a poster at the annual congress of the Belgian Society for Internal Medicine (BSIM) in 2018.

Statement of ethics

Written informed consent was obtained for anonymous publication of the data presented here.

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