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Review

A review of 42 cases of intestinal pseudo-obstruction in patients with systemic lupus erythematosus based on case reports

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ABSTRACT

Intestinal pseudo-obstruction (IpsO) is considered a severe manifestation of systemic lupus erythematosus (SLE) characterized by clinical and radiological evidence of intestinal obstruction with no identifiable mechanical lesion. We performed a systematic review to document IpsO in SLE. Twenty-eight articles with 42 patients were included. The median age of onset of IpsO was 27.5 (10–57) years. The female to male ratio was 38:4. Twenty-two (52.4%) patients had IpsO as the initial presentation of their underlying lupus. Three (7.1%) patients manifested in inactive lupus. The duration of abdominal symptoms before admitted ranged from 3 days to 3 years, however most of the patients responded well to systemic corticosteroid or immunosuppressive treatment within 2 days to about 3 months. Concomitant ureterohydronephrosis was present in approximately three-fourths of the cases. More interestingly, 4 patients presented hepatobiliary dilatation without mechanical obstruction together with IPO and ureterohydronephrosis. In conclusion, IpsO is an uncommon but important manifestation of SLE. The finding of coexisting ureterohydronephrosis and hepatobiliary dilatation suggests that there may be generalized visceral muscle dysmotility. Early recognition of IpsO is necessary to institute appropriate medical treatment and to avoid inappropriate surgical intervention.

Contents

Abbreviations: IpsO, intestinal pseudo-obstruction; SLE, systemic lupus erythematosus; ACR, American College of Rheumatology; CTD, connective tissue disease; GML, generalized megaviscera of lupus; VMDS, visceral muscle dysmotility syndrome.

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1. Introduction

SLE is a multisystemic autoimmune inflammatory disease with a variety of presenting features and manifestations. SLE can involve any part of the gastrointestinal tract with oral lesions, esophageal dysmotility, mesenteric vasculitis, protein-losing enteropathy and pancreatitis the most frequent manifestations [1,2]. Gastrointestinal symptoms are common in patients with SLE, with more than half of them complaining of anorexia, nausea and vomiting [1]. The symptoms may be directly attributable to SLE, intercurrent illness or side-effects of medication [1]. Abdominal complaints in patients with SLE may pose a difficult diagnostic challenge with the possible differential diagnoses of primary gastrointestinal disorders, manifestations of SLE on the gastrointestinal tract and side effects of therapy for SLE. IpsO is a rare clinical syndrome characterized by ineffective intestinal propulsion with signs and symptoms similar to mechanical bowel obstruction including abdominal distension, pain, nausea, vomiting, constipation and hypoactive or absent bowel sounds, but the absence of an identifiable organic obstructive lesion [3]. IpsO reflects a dysfunction of the visceral smooth muscle or the enteric nervous system or the visceral autonomic nervous system or other unclear pathological changes. IpsO may be primary or secondary to an underlying systemic disease. Secondary causes include neurologic, endocrine, and connective tissue disease (CTD) [3,9,25]. Among CTD, systemic sclerosis is the most common underlying disease. Secondary IpsO has been recognized as an uncommon manifestation of SLE in recent decades [4–29]. It can manifest as a complication of the disease or, less commonly, as the initial presentation [25,28,30]. It usually occurs during active lupus but may manifest itself in inactive lupus [24]. It seems that most patients had a satisfactory response to corticosteroids or immunosuppressants and the diseases can be potentially reversible with conservative treatment in an early stage, but in some cases this complication evolved regardless of the underlying disease activity [3,23–25,28,30]. Physician awareness and early diagnosis could positively affect outcomes of this syndrome, avoiding unnecessary surgical intervention.

There have been 42 cases of IpsO associated with SLE to date in the English literature. The clinical profile, immunological features, histological findings, treatment modality and disease outcome of these patients are reviewed. Possible underlying mechanisms of this complication are discussed.

2. Methodology

An English literature search revealed 28 articles with 42 patients satisfying the American College of Rheumatology (ACR) revised criteria for the classification of SLE [31] who had IPO as a clinical manifestation of their underlying lupus. Patients fulfilling less than 4 of the ACR criteria were not included in our revision, although the diagnosis of SLE was highly suggestive. The disease activity in SLE was assessed according to the most widely used criteria of ECLAM or SLEDAI [32]. The review was made using the MedLine PubMed database from 1966 to the present. Subject terms used for the search were intestinal pseudo-obstruction and systemic lupus erythematosus. We also reviewed reference lists of included studies, systematic reviews and the online publication lists of highly referenced studies to identify potentially relevant studies that may not have been identified in our literature searches.

3. Results

3.1. General clinical characteristics

The characteristics of all 42 cases are summarized in Table 1. The female to male ratio was 38:4. Twenty-two (52.4%) patients had IpsO as the initial presentation of their underlying lupus. Three (7.1%) patients manifested as inactive lupus. The duration of abdominal symptoms before admitted ranged from 3 days to 3 years, however most of the patients responded well to systemic corticosteroid or immunosuppressive treatment within 2 days to about 3 months. The small bowel was more frequently involved than other parts of the gastrointestinal tract. Nine patients underwent surgical intervention because of symptoms of intestinal obstruction.

3.2. An apparent association with ureterohydronephrosis

Concomitant organ involvement by the lupus process during IpsO (Table 2) was more subtle and required a high level of clinical awareness for diagnosis. Concomitant ureterohydronephrosis was present in approximately three-fourths of the cases. It is noteworthy that 4 patients presented hepatobiliary dilatation without mechanical obstruction together with IpsO and ureterohydronephrosis.

3.3. Serological data

The serological data and autoantibody profile of the previously reported patients was incomplete. No specific autoantibodies in IpsO were found. The detailed information on SLE, including laboratory indices and other organ involvement were listed in Table 3.

3.4. Disease outcome

There was a 9.5% (4/42) mortality in the series reviewed, three within 1 year of presentation of IpsO. The causes of death were mainly related to infections (2/4) and other major organ involvement (3/4) of SLE, especially progressive renal failure and cerebromeningeal haemorrhage. Eight patients (19.0%, 8/42) had an undulant course with recurrent attacks of IpsO. The follow-up rate is low, so the long-term outcome of patients with IpsO is unclear.

4. Discussion

Systemic lupus erythematosus is the most prevalent autoimmune disease, with an annual incidence of 60 per 1 million population and a prevalence of 500 per 1 million population [33]. Because of its myriad of multisystemic clinical presentations, systemic lupus erythematosus is known as one of the great medical mimics alongside tuberculosis, syphilis, and human immunodeficiency virus. Gastrointestinal complications have been increasingly observed as manifestations of SLE, and occur in 50% of them at any time during the course of their illness, but only 2–30% of these are directly attributable to SLE, the rest being side effects of the medication or intercurrent diseases [1,2].

IpsO is a rare clinical syndrome characterized by ineffective intestinal propulsion with clinical and radiological evidence of intestinal obstruction, but the absence of an identifiable mechanical lesion [3]. It can be either idiopathic or secondary to an underlying disease. IpsO is idiopathic in most cases. Secondary causes include neurologic, endocrine, and connective tissue disease (CTD) [3,9,25]. Among CTD, systemic sclerosis is the most common underlying disease. However, there are only a few case series describing SLE-related IpsO. So we performed the systematic review to document the characteristics of IpsO as a clinical manifestation of SLE; thus help to institute appropriate medical treatment and avoid inappropriate surgical intervention.

Recently, IpsO has been recognized as an uncommon and poorly understood complication of SLE when Cacoub et al. [6] described the first proven case in a patient presenting a severe form of SLE