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Case Report

CASE REPORT

Challenging case of ischemic colitis, necrotic cutaneous vasculitis and thromboembolic disease in an elderly patient with Sjogren's syndrome

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Abstract

Sjogren's syndrome is a chronic autoimmune condition characterized by reduced lacrimal and salivary gland secretions. In a minority of the cases, patients can develop rarer complications, such as vasculitis and, even less commonly, ischemic colitis. Herein, we present a challenging case of a 73-year-old woman with a background of Sjogren's syndrome (SS) who initially presented with a purpuric rash on the right leg. She was initially managed with antibiotics and referred for an outpatient rheumatology review. A few days later, she was readmitted to the hospital generally unwell with a widespread rash. She developed deep vein thrombosis and per rectal bleeding secondary to ischemic colitis. She had excellent response to medical management including steroid therapy and azathioprine. This case highlights the very rare complications of SS. Whilecutaneous vasculitis is not uncommon in primary Sjogren's, ischemic colitis is very rare and is a potentially serious complication, which requires prompt diagnosis and management.

INTRODUCTION

Sjogren's syndrome (SS) is a chronic autoimmune condition characterized by reduced lacrimal and salivary gland secretions. In a minority of cases, patients can develop rarer complications, such as vasculitis and, even less commonly, ischemic colitis.

Herein, we present a challenging case of a 73-year-old woman with background of SS who initially presented with a progressive purpuric rash on the right leg. She had rapid deterioration with a widespread vasculitic necrotic rash. She also has deep vein thrombosis (DVT) and per rectal bleeding secondary to ischemic colitis.

CASE REPORT

This is a case of a 73-year-old woman who was originally admitted to the acute medical unit with a 6-week history of progressive right leg swelling and purpuric rash. During this period she had been initially managed with numerous courses of antibiotics in the community; while the swelling improved, the rash did not.

Other history of note included generalized myalgia, lethargy and longstanding shortness of breath on exertion.

She has a medical history of Anti RO/LA-positive SS diagnosed 10 years ago, angina, hypertension, hypercholesterolemia, peripheral neuropathy, hypothyroidism, mitral valve dysfunction and iron deficiency anemia.

Her long-term medication include the following: ivabradine, isosorbide mononitrate, clopidogrel, amitriptyline and levothyroxin.

On initial assessment, she was systemically well, afebrile and had evidence of a mild purpuric rash on both ankles with background erythema and swelling of the left ankle. The remainder of the physical examination was unremarkable.

The inflammatory markers were raised with a C-Reactive protein (CRP) of 20 mg/l and a White cell count (WBC) count of 15

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(10^3/mm^3) with a neutrophilia of 10 (10^3/mm^3). Initial management was with further antibiotics, and she was discharged with outpatient rheumatology follow-up planned.

However, a week later she was readmitted feeling generally unwell with worsening vasculitic rash extending to both the upper and lower limbs with associated necrotic changes on her fingertips. She rapidly deteriorated and became septic. She also developed episodic rectal bleeding associated with abdominal pain and described lower left limb pain and swelling. Clinical examination was suspicious of DVT, and her abdomen was mildly distended and generally tender.

The ultrasound Doppler of the lower limbs confirmed extensive thrombus of the left common femoral vein, superficial femoral and popliteal veins. A computed tomography scan of the abdomen revealed marked mucosal thickening of the bowel from the splenic flexture to the rectum associated to pericolic inflammatory changes in keeping with acute colitis. As the patient was allergic to omnipaque, intravenous (IV) contrast was not used. Flexible sigmoidoscopy and biopsy confirmed the diagnosis of ischemic colitis.

Medical management with IV fluid resuscitation, antibiotics and heparin for confirmed DVT was initiated. Following rheumatology review, she was treated with IV methylprednisolone, 500 mg for 1 hour for 3 consecutive days and oral prednisolone 1 mg/kg per day for induction of clinical remission. We also started her on azathioprine for further management of her vasculitis.

DISCUSSION

SS is an autoimmune exocrinopathy predominantly affecting the lacrimal and the parotid glands thus resulting in classical sicca syndrome [1]. It can also affect other organs with skin vasculitis reported as one of the most common extra glandular manifestations, particularly in Anti RO/LA-positive subtypes, which tend to have higher rate of systemic manifestations [2]. A large study examining SS clinical features in over 1000 primary SS patients demonstrated that vasculitis also increased among those with a long duration (>10 years) of the disease [2].

SS-associated vasculitis affects mainly small and medium vessels and can vary from petechial to extensive purpuric necrotic rash as was seen in this case [3]. It can also have complications including intestinal involvement, and it can also increase the risk of thromboembolic disease as we report.

To the best of our knowledge, no similar cases with concomitant extensive skin vasculitis, ischemic colitis and thromboembolism in patients with SS are described in the literature.

Thromboembolic disease is a recognized complication of SS [4]. SS can also involve any part of the gastrointestinal system including the small and large bowel. Furthermore, SS can be associated with inflammatory bowel disease, Crohn's disease or ulcerative colitis [5]. Ischemic colitis can be due to atherosclerosis or autoimmune secondary to mesenteric inflammatory veno-occlusive disease and has been reported to affect the colon in more than 50% of the cases [5,6]. It is characterized by mesenteric venous inflammation and thrombosis resulting in bowel ischemia.

This condition represents a diagnostic and management challenge particularly in elderly patients with complex comorbidities.

Supportive medical treatment in most cases can help to resolve symptoms with immunosuppressive treatment (as used in this case) can make a significant difference to patient outcomes.

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CONFLICT OF INTEREST STATEMENT

None declared.

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ETHICAL APPROVAL

This case report does not require ethical approval. However, it does meet the ethical guidelines. An informed consent was obtained from the patient, and confidentiality was maintained throughout the process.

CONSENT

An informed written consent was obtained from the patient.

GUARANTOR

There no guarantor to declare.

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