

Ascites as the Presenting Symptom in a Patient with Churg-Strauss Syndrome

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Abstract

A 67-year-old male presented with ascites, dyspnoea, peripheral edema and purpura. The history revealed asthma and nasal polyps. The laboratory tests showed an increased peripheral blood eosinophilic cell count. The ascitic fluid analysis showed features consistent with an eosinophilic peritonitis. A skin biopsy revealed eosinophilic vasculitis. Our patient was diagnosed with Churg-Strauss syndrome based on the medical history, laboratory and histology. This case describes ascites as the presenting symptom of Churg-Strauss syndrome.

Key words

Churg-Strauss syndrome – ascites – purpura – vasculitis – eosinophilic peritonitis.

Introduction

Churg-Strauss syndrome (CSS) is a clinical syndrome characterized by asthma, hypereosinophilia and systemic necrotizing vasculitis and/or extravascular eosinophilic granulomas. It is a rare systemic autoimmune disease with an estimated incidence of 2.4 per million per year in the general population [1-4].

Most patients have a history of asthma and allergic rhinitis. Some authors suggest that there are three phases of CSS: 1) a prodromal phase with allergic rhinitis, nasal polyps and/or sinusitis followed by asthma which can present many years before manifestation; 2) an eosinophilic phase with features of peripheral blood eosinophilia and eosinophilic infiltration of multiple organs, and eventually 3) the vasculitic phase with symptoms of systemic vasculitis. In the latter phase the diagnosis of CSS can be made [2, 5, 6].

Every organ may be affected. Most commonly involved are the lungs, skin, peripheral nerves, heart and gastrointestinal tract. Eosinophilic infiltration of the gastrointestinal tract often presents with abdominal pain and bloody diarrhoea [1-5]. Eosinophilic peritonitis and ascites however is rarely described in literature [1, 10]. This patient had ascites as the presenting symptom of CSS.

Case report

A 67-year old male was referred to our hospital because of unexplained ascites, progressive dyspnoea and purpura. His medical history revealed asthma and surgical treatment for recurrent nasal polyps. He complained of slowly progressive dyspnoea and general malaise. He had lost more than thirty pounds, despite increased abdominal circumference and peripheral edema.

On admission, the patient was moderately dyspnoeic. His temperature was 36.2°C, blood pressure 96/63 mmHg, pulse 91 beats per minute. No pathological lymph nodes were found. Auscultation of the lungs revealed expiratory wheezing. There were normal heart sounds. The abdomen was distended, nontender with normal bowel sounds and shifting dullness. The liver edge did not descend below the costal margin and the spleen had normal size. Extensive peripheral edema and palpable purpura were found on both hands, legs and flanks. Neurological examination was consistent with symmetrical sensory peripheral neuropathy.

Laboratory tests showed an increased erythrocyte sedimentation rate and increased peripheral blood eosinophil count (Table I). The ascitic fluid analysis showed an exsudate, containing increased number of eosinophils. Chest X-ray demonstrated left- and right-sided pleural effusions. A computed tomography (CT) scan of the chest to exclude pulmonary embolism was negative. Pleural fluid analysis findings were similar to those of ascites: exsudate containing increased number of eosinophils. Lung function testing was performed and showed decreased forced expiratory volume in one second (FEV1) compatible with broncho-obstructive disease.

Due to the differential diagnosis of an enteropathy

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Table I. Laboratory results in our patient

Tests	Results	Normal
Erythrocyte sedimentation rate (mm/hour)	74	1-25
C reactive protein (mg/L)	45	0-10
Hemoglobin (mmol/L)	7.6	7.5-10.0
Leukocytes (x 10 ⁹ /L)	7.0	4.0-10.0
Eosinophils absolute (x 10 ⁹ /L)	0.56	0.0-0.5
Albumine (g/L)	17	35-55
NT-proBNP (Ng/L)	185	0-738
Sodium (mmol/L)	135	135-145
Potassium (mmol/L)	5.1	3.5-5.0
Creatinine (μmol/L)	102	50-100
Urea (mmol/L)	6.0	2.5-7.5
ANCA	Doubtfull positive	

associated lymphoma, an anterograde double balloon enteroscopy with random biopsies was performed revealing gastric intraepithelial lymphocytosis (mainly plasma cells). An additional skin biopsy was compatible with eosinophilic vasculitis (Fig. 1).

The diagnosis of CSS was made according to the criteria of the American College of Rheumatology (ACR): the presence of a history of recurrent nasal polyps and asthma, sensory neuropathy, elevated peripheral blood eosinophile count, extravascular eosinophilic infiltration and vasculitis in biopsy specimens [11].

Treatment with high-dose prednisolone and azathioprine was instituted and the ascites, skin lesions and dyspnoea subsided within a week. Almost a year after diagnosis and initiation of treatment, he is in complete remission whilst using prednisolone 10 mg OD and azathioprine 50 TDS.

Discussion

Our patient presented with ascites, malaise and progressive dyspnoea. The differential diagnosis of ascites is extensive and mainly includes liver cirrhosis, congestive heart disease, nephrotic syndrome, lymphoma and other malignancies or infectious and inflammatory diseases such as tuberculosis, pancreatitis, constrictive pericarditis, sarcoidosis, Whipple's disease and celiac sprue. However, in textbooks and other medical sources CSS is seldom mentioned in this differential diagnosis.

Churg-Strauss syndrome is a systemic small vessel vasculitis characterized by eosinophilia, granulomatosis and vasculitis. Most patients present with general features of malaise, fever and weight loss. Churg-Strauss syndrome can affect any organ, but most frequently the respiratory tract is involved. More than 95 percent of patients suffer from asthma and most patients have patchy transient pulmonary infiltrates. Pleural effusions arise occasionally. Peripheral neuropathy and skin lesions are also common [1, 4, 7].

Gastrointestinal manifestations as the presenting symptom are seldom described [7]. In most patients, CSS develops in three stages. In the first stage asthma develops.

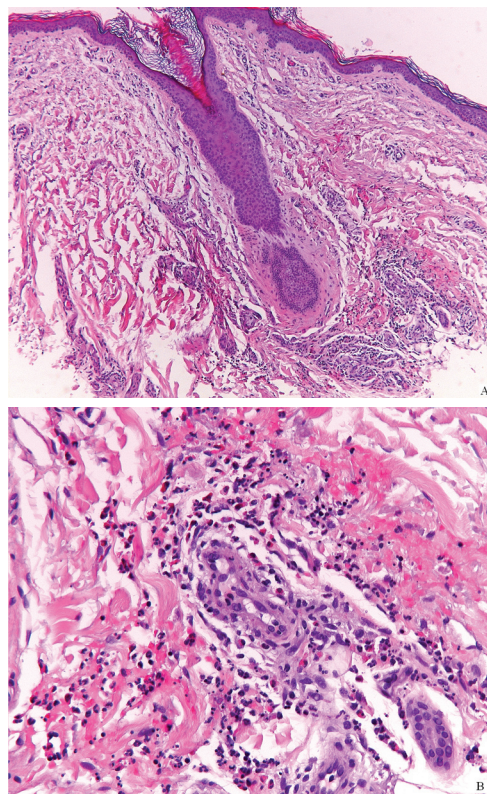


Fig 1. The images show an overview (A) (x10) and detail (B) (x20) of the skin biopsy stained with H&E in which a dense eosinophilic infiltrate around some small vessels can be seen at the basis of a hair follicle, with deposition of some nuclear debris. The endothelium has slightly plumb nuclei, but there is no fibrinoid necrosis. There is extravasation of some erythrocytes, indicative of vascular leakage.

In the second stage infiltration of tissue by eosinophilic cells occurs. In this stage differentiation from other hypereosinophilic conditions - eosinophilic gastroenteritis and idiopathic hypereosinophilic syndrome - can be difficult. The third stage is characterized by the onset of systemic vasculitis and usually develops several years after the onset of asthma. In this stage, features consistent with pancreatitis, mucosal colonic ulcers, gastrointestinal perforation, or bloody diarrhoea should arise the suspicion of CSS [1- 6, 8, 9]. Random colonic biopsies will reveal necrotizing vasculitis of medium-to-small sized blood vessels and eosinophilic infiltration of the vessels and perivascular tissues, sometimes even with granuloma formation [7]. However, ascites as the presenting symptom of CSS is never described.

Suen and Burton described a classification of gastrointestinal involvement of patients with hypereosinophilic syndromes into three categories: 1) predominant mucosal disease with signs of iron deficiency anemia, hypoalbuminemia and steatorrhea; 2) predominant muscle layer disease resulting in obstructive symptoms and 3) predominant subserosal disease associated with peritonitis and ascites [10]. However, no other reports have been published regarding the association of CSS and eosinophilic peritonitis. This is the first reported case of CSS presenting with ascites.

In **conclusion**, when a patient presents with ascites and a remarkable history of nasal polyps and asthma, the diagnosis of CSS should be considered in the differential diagnosis.

Conflicts of interest

The authors have no conflicts of interest or financial disclosures to report.

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