Churg-Strauss abdominal manifestation

ABSTRACT

Background: Churg-Strauss is a rare, idiopathic, hypereosinophilic disease characterized by blood, tissue, and systemic vasculitis in patients with a history of asthma or allergic rhinitis. Gastrointestinal manifestations of Churg-Strauss appear in 31-45% of patients according to some studies, abdominal pain being the most frequent symptom followed by diarrhea and bleeding.

Clinical case: We present the case of a male patient with a history of asthma who presented abdominal pain apparently due to acute appendicitis. During the hospital stay, the study protocol confirmed diagnosis of Churg Strauss syndrome with intestinal manifestations.

Conclusion: Churg Strauss syndrome is a rare form of vasculitis that may present with intestinal manifestations. It is therefore important to take into consideration the differential diagnosis. Few cases have been reported in the literature associated with this syndrome and acute abdomen, all reporting a poor prognosis of this association.

Key words: Churg Strauss syndrome, necrotizing vasculitis, intestinal vasculitis.

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BACKGROUND

Allergic granulomatosis, angiitis and necrotizing vasculitis with peripheral eosinophilia (or Churg-Strauss Syndrome) is a rare idiopathic disease characterized by hypereosinophilia in blood and tissues coupled with systemic vasculitis in patients with a history of asthma or allergic rhinitis.¹

The classic lesion described by Churg and Strauss is allergic granuloma, although its absence does not exclude the diagnosis.² Churg-Strauss syndrome manifests itself with a wide range of symptoms where asthma affects almost all individuals (97%) and can precede vasculitis even by 10 years; 61% of patients will have sinusitis, 40% arthralgias and 49% skin lesions.³ Gastrointestinal manifestations of Churg-Strauss syndrome are demonstrated in 31-45% of patients according to some studies. Abdominal pain is the most reported symptom followed by diarrhea and hemorrhage. Surgical intervention is rarely required; however, intestinal inflammation can cause peritoneal irritation with manifestations of acute abdomen. Dehiscence of the repaired tissues is an additional complication in surgical patients and reflects the nutritional state, steroid dose and systemic inflammatory effect of the underlying vasculitis.⁴ The objective of this study is to report the clinical case of a patient with intestinal vasculitis and to review the literature.

CLINICAL CASE

We report the case of a 27-year-old male patient with a history of asthma treated with steroids and bronchodilators since childhood. One month prior to hospital admission he noticed petechiae in the bilateral inguinal region and increase in testicular volume, which progressively disappeared with medical treatment. Upon admission to the hospital he reported abdominal pain localized in the right iliac fossa, 9/10 in intensity, accompanied by nausea, vomiting of biliary content, diarrhea with cramping, and tenesmus.

On physical examination the patient was diaphoretic, tachycardic at 100 and tachypneic 25/min, bibasilar pulmonary hypoventilation, distended abdomen, pain on palpation of the iliac fossa (right predominance) and McBurney’s point, and questionable Von Blumberg sign.

Chest x-ray showed grade III cardiomegaly, parenchymal hyperlucency, flattening of the right hemidiaphragm, straightening of the costal arches, increase in the bronchiovascular connection and diffuse pulmonary infiltrates (Figure 1). Abdominal x-rays showed air fluid levels in the right iliac fossa and right psoas effacement (Figure 2). Blood count showed leukocytosis of 26,000 cells/mm³, 58% neutrophils, 17% lymphocytes, and 23% eosinophils. Ultrasound showed an unquantified right pleural effusion, gallbladder with double hale and thickened wall. In the right iliac fossa there was a fixed plastron dependent on the intestinal loops. Chest tomography confirmed a moderate bilateral pleural effusion and pericardial effusion. Abdominal tomography corroborated a tumor in the right iliac fossa dependent on the cecum with luminal stenosis (Figure 3). On exploratory laparotomy there were findings of ischemia in the cecum and terminal ileum and free fluid in the peritoneal cavity. A right hemicolectomy with transverse ileum and anastomosis was carried out. During the postoperative period, the patient underwent supplemented studies due to the appearance of petechiae in the extremities (Figure 4). Echocardiogram reported overall pericardial effusion of 1200 cc with no evidence of tamponade and ejection fraction of 67%. According to the immunological profile, IgE concentrations of 5 and 125 and IgG 1028 were notable with the remainder being negative. On the third postoperative day the patient was in a state of shock and had data of acute abdomen. For this reason, a second exploratory laparotomy was carried out, finding dehiscence of the anastomosis. An ileostomy and distal closure of the Hartmann pouch was done.
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**Figure 1.** Radiograph of the chest showing cardiomegaly.

**Figure 2.** Abdominal x-ray with multiple air fluid levels towards to the right iliac fossa.

**Figure 3.** Abdominal tomography where a tumor is observed in the right iliac fossa dependent on the cecum.

**Figure 4.** Multiple petechiae in the lower extremities.

Histopathological report of the specimen was panmural acute ischemic colitis with thrombosis in the small- and medium-sized vessels that mainly affected the cecum wall, the mucosa of the ascending colon and terminal ileum with unspecified acute and chronic inflammation, vascular congestion and focal thrombosis of the small- and medium-sized vessels.

The patient's subsequent hospital evolution was towards improvement, hemodynamic stability, and oral feeding tolerance with functional stoma. The patient was discharged from the hospital and
subsequent treatment was in conjunction with physicians from the rheumatology service on an outpatient basis.

Four months later there was a notable improvement in the nutritional status. Intestinal transit was reestablished without complications. The patient remains in stable condition and is followed as an outpatient.

**DISCUSSION**

In 1950 Churg and Strauss published this syndrome in a series of autopsies of patients with severe asthma accompanied by fever, hypereosinophilia, symptoms of renal and heart failure and peripheral neuropathy secondary to systemic vasculitis. Churg-Strauss syndrome is a necrotizing vasculitis of small vessels (≤ 0.5 mm). The mean age of appearance is 30 years. Abdominal pain is present in 45% of the cases with unknown cause.

In 1990 the American College of Rheumatology proposed six criteria to classify the disease. To establish the diagnosis, 4/6 are needed with a sensitivity of 85% and specificity of 99.7%: 1) asthma, 2) eosinophilia > 10% differential, 3) mono- or polynuropathy, 4) diffuse pulmonary infiltrates on x-rays, 5) abnormalities in the paranasal sinuses, 6) biopsy with blood vessels with extravascular eosinophils. In this case, four of the criteria mentioned were present.

Mesenteric vasculitis is a rare cause of intestinal ischemia found in only 2% of the cases, almost always associated with lupus, rheumatoid arthritis, giant cell arteritis, Wegener granulomatosis and Churg-Strauss syndrome.

Among the gastrointestinal manifestations are acute abdominal pain, bloody bowel movements, diarrhea and, occasionally, nausea and vomiting. The small intestine is the most affected.

Our patient had a clinical picture of acute appendicitis; however, during surgery necrosis was found in the cecum and the appendix. Ischemia was secondary to mesenteric vasculitis. These patients may also have hemorrhage or perforation. In patients with Churg-Strauss syndrome, angiography shows microaneurysms, stenosis and rarely thrombosed arteries.

Gastrointestinal disorder is the leading cause of death in Churg-Strauss syndrome. Abdominal pain is not associated with a poor prognosis, but severe gastrointestinal disease has a poor prognosis (bleeding, perforation, infarction or pancreatitis). The rate of survival in patients with gastrointestinal disease can even reach 43.2% at 6 years compared with patients without the disorder (78.9%). Mortality in patients with acute abdomen is 44% compared with those who do not have acute abdomen (18%).

Churg and Strauss found myocardial abnormalities in > 50% of the autopsies, ranging from extensive substitution of the myocardium by granulomas and scar tissue to vasculitis in the coronary vessels.

Myocardial damage may be caused by toxic mediators related to activated eosinophilic infiltrates or to lesions in the myocardium and vasculitis in the coronary vessels. Myocarditis can lead to postinflammatory fibrosis and to restrictive myocardial disease or congestive heart failure, whereas coronary vasculitis can give rise to ischemic heart disease.

Acute intestinal ischemia is a surgical emergency with life-threatening consequences. Mesenteric vasculitis is a rare cause of intestinal ischemia and represents 2% of cases. Churg-Strauss syndrome together with rheumatoid arthritis, scleroderma, systemic lupus erythematosus, giant cell arteritis, and Wegener granulomatosis are systemic diseases that on rare occasions may cause ischemia or intestinal infarction.
It is an infrequent disease and after diagnostic confirmation it is important to treat each of its clinical manifestations. In the case of operated patients the possible risks and complications should be kept in mind. The histopathological report of the surgical specimen is mandatory to confirm diagnosis.

In conclusion, Churg-Strauss syndrome is an uncommon vasculitis that could manifest itself with intestinal symptoms such as the case reported here. It should be kept in mind as part of the differential diagnoses. The immunological status of the patient is relevant for making decisions in cases of intestinal perforations due to the high risk of dehiscence. There are few cases in the literature associated with this syndrome and acute abdomen and all are focused on the poor prognosis of this association.

REFERENCES