Gastrointestinal Manifestations of Churg-Strauss Syndrome

Ellen C. Ebert
UMDNJ-Robert Wood Johnson Medical School, New Brunswick, NJ 08903

Abstract

Background and Aims: The diagnosis of Churg-Strauss syndrome (CSS), an allergic granulomatosis involving small- and medium-sized vessels, requires the presence of at least four of six criteria including asthma, peripheral eosinophilia, and systemic vasculitis. The gastrointestinal manifestations have never been reviewed.

Methods: The 75 articles in the English literature found by electronic searches from 1960 to 2011 were examined.

Results: Ischemia from vasculitis causes ulcerations, perforation, annular stenosis, and/or intestinal occlusions usually involving the small bowel, presenting as an acute abdomen or intestinal angina. Colonoscopy characteristically shows shallow ulcers with erythematous halos. Endoscopic biopsies usually do not reveal vasculitis as this requires sampling of the submucosa. Intestinal resections reveal eosinophilic infiltrates, vasculitis, fibrinoid necrosis, and/or granulomas. Unusual complications include acalculous cholecystitis and eosinophilic ascites from involvement of the peritoneum. Most patients respond to corticosteroids although mesenteric ischemia is associated with a poor outcome.

Conclusion: CSS, while rare, has characteristic GI involvement.

Keywords: Vasculitis; Ischemia; Ulcerations; Cholecystitis

Introduction

Churg Strauss syndrome (CSS) was first defined in 1951 as an allergic granulomatosis affecting small and medium-sized vessels [1]. The American College of Rheumatology established six criteria for its classification in 1990: asthma, peripheral eosinophilia, sinus involvement, pulmonary infiltrates, mononeuritis multiplex, and histology compatible with vasculitis [2]. The presence of 4 or more criteria yielded a sensitivity of 85% and a specificity of 99.7%. Before that, three criteria—asthma, peripheral eosinophilia, and systemic vasculitis—were needed [3]. Histological lesions include small-vessel angiitis and extravascular necrotizing granulomas, usually containing eosinophilic infiltrates [4]. Vasculitis may be granulomatous or nongranulomatous and typically involves both arteries and veins in pulmonary and systemic vessels [5]. The granulomas have a center formed by necrotic eosinophils surrounded by palisading histiocytes and multinucleated giant cells [4]. They are not present in all patients and are seen in other diseases. There are three partially-overlapping phases in CSS [4]: 1) a prodromal phase with allergic rhinitis, nasal polyps, and/or sinusitis, followed by asthma; 2) an eosinophilic phase with peripheral blood eosinophilia and eosinophilic infiltration of organs, and 3) a vasculitic phase.

This is a review of the GI and hepatic manifestations of CSS using the 75 articles in the English literature from 1960 to 2011 found using a Medline search, Google Scholar, and the references in the articles obtained. The emphasis is on recent literature, particularly studies fulfilling the 1990 criteria for diagnosis, although the description of cases is not always adequate to make this determination.

GI involvement occurs in 17% to 59% of cases [3,6]. Of those with GI disease, abdominal pain is present in 22% to 97% of patients, diarrhea in 9% to 33%, hematochezia or melena in 6% to 18%, and a surgical abdomen in 9% to 34% [3,7-9]. The manifestations are diverse including features of both eosinophilic gastroenteritis and polyarteritis nodosa (PAN). CSS is differentiated from eosinophilic gastroenteritis by the presence of vasculitis. Compared to CSS, PAN less commonly has pulmonary involvement but is more commonly associated with microaneurysms, bowel vasculitis, relapses, and death [10].

Involvement of the upper GI tract

There may be oral aphthous lesions and palatine ulcers in CSS [11]. “Congestive” esophagitis as an initial manifestation of disease has been described with markedly thickened esophageal wall presenting as odynophagia or abdominal pain with nausea and vomiting [12]. Esophageal perforation may occur [9]. Ulcers in the stomach and duodenum with eosinophilic infiltration and thickened vessel walls may resolve with corticosteroid therapy [13,14]. Also described are gastric serosal plaques and subserosal nodules [13]. Diffuse thickening, distortion, and obliteration of the gastric rugal folds with intramural masses responding to immunosuppression may occur [15].

Involvement of lower GI tract

Ischemia from vasculitis causes ulcerations, perforations, anular stenosis, and/or intestinal occlusions, usually involving the small bowel, presenting as an acute abdomen or intestinal angina [16,17]. Perforations are said to be more common in the Japanese than the English literature, usually in patients already on corticosteroids [18]. However, some of the case studies available in complete [18] or abstract form [19,20] have less than four criteria for the diagnosis. CSS may be associated with Crohn’s disease [21]; whether this is a true association or a chance co-existence of two diseases is unknown. As in all immunocompromised hosts, infectious enteritis, as with cytomegalovirus, must be kept in mind [22]. Capsule endoscopy may be useful in the diagnosis of small bowel lesions [23]. Angiography sometimes shows aneurysms that may rupture [24].

Colonic involvement presents with abdominal pain, bleeding, and diarrhea. Perforated appendicitis from vasculitis may occur [7], rarely associated with enteroenteral fistulas [25]. Most commonly

Corresponding author: Ellen C. Ebert, UMDNJ-Robert Wood Johnson Medical School, New Brunswick, NJ 08903, Tel: 732-235-7784; Fax: 732-235-7792; E-mail: jeydeils@yahoo.com

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described are shallow ulcers with erythematous halos, although not all of these cases had four criteria for the diagnosis [26-30]. Also found are subepithelial hemorrhages [31] or pseudopolyps suggesting ulcerative colitis [6]. An allergic granulomatous process massively involving the ascending colon mimicked carcinoma [6].

Vasculitis is more often shown on intestinal resections than on colonoscopic biopsies, where the histologic diagnosis of CSS was made in only 3 of 11 cases [28]. GI biopsies rarely exhibit vasculitis unless they sample deep in the submucosa, which can be hazardous, especially in patients with ischemic GI involvement [32]. Histology may show an eosinophilic infiltrate, nonspecific vasculitis, ischemic changes, fibrinoid degeneration and luminal narrowing, leukocytoclastic capillaritis and eosinophilic vasculitis, granulomatous and necrotizing vasculitis, and eosinophil crypt abscesses [16,27,28,30,31,33-35].

Involvement of liver, gallbladder, pancreas, and omentum

Hepatomegaly occurs in 9% and abnormal liver tests in 6% to 7% of patients with CSS [8,9], sometimes due to vasculitis with eosinophilic infiltrates or to cyclophosphamide therapy [21]. Necrotizing granulomas with infiltrates of eosinophils and hepatomegalias responding to corticosteroids have been described [36]. Periporal hepatitis with an eosinophilic infiltrate developed into a picture consistent with primary biliary cirrhosis [37]. Abdominal distension due to eosinophilic ascites, suggesting involvement of the peritoneum, sometimes responds to immunosuppression [32,33,38]. Also described are hepatic artery aneurysms and infarcts [3], with an aneurysm rupturing into the intrahepatic duct causing melena [39]. Budd-Chiari syndrome or hepatic venous outflow obstruction has been described, responding to anticoagulation and/or immunosuppression [40,41].

Acatalolic cholecytitis may be associated with an eosinophilic inflammation, small vessel vasculitis, granulomas, necrotizing panarteritis, and/or fibrinoid necrosis [33,42-44]. A necrotizing or suppurrative, perforated cholecystitis may develop [7,21]. Nodules containing epithelioid granulomas with multinucleated giant cells have been seen [45]. Acute pancreatitis may develop [9].

Omental nodules with an allergic granulomatous angiitis involving the veins with segmental lesions at different stages of development [32] and an omental hematoma [9] have been described. Omental resection in another case showed endoluminal thrombosis and a chronic inflammatory infiltrate of the small-sized mesenteric vessel walls [21]. Serosal nodules, localized or disseminated, may be scattered all along the gut [32].

Treatment and prognosis

Corticosteroids have greatly improved the prognosis of CSS with about 90% achieving remission [9,46]. Other immunosuppressive agents, such as cyclophosphamide, can be added if symptoms are not controlled. Many GI lesions respond to corticosteroids [14-15,21,24,28,32,38,40-41]. A successful identical-twin living donor small bowel transplant for necrotizing vasculitis has been reported [47].

According to multivariate analysis, myocardial involvement and severe GI disease secondary to mesenteric ischemia are significantly associated with a poor clinical outcome [9]. Bowel infarction accounted for 27% of deaths in one series.

References


