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Title page

BOWEL PERFORATIONS IN A PATIENT AFFECTED BY CHURG-STRAUSS SYNDROME
UNDER HIGH DOSE STEROID TREATMENT: MAY ALTERNATIVE DRUGS REDUCE RISK OF
SURGERY?

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Cover letter: this essay wants to add to literature one more case of large bowel ischemia and perforation in a patient affected by Churg-Strauss Syndrome. I reviewed the previous reports of intestinal perforation in patients affected by this disease. Up to now there are no reports about the effect of steroids on this complication and the use of alternative drugs in order to reduce it. We underline the carefully use of steroids in gastro intestinal involvement of acute Churg-Strauss Syndrome and the possible advantages of alternative drugs.

ABSTRACT

Churg Strauss Syndrome is a relapsing-remitting vasculites that frequently involves digestive system. Ischemic perforation of the large bowel is relatively rare and potentially life threatening. We report a case treated with high dose of steroids for a relapsing of Churg-Strauss vasculopathy that underwent emergency surgery for multiple large bowel's perforations. Massive use of steroids is common to control relapsing of this disease but it increase the risk of intestinal perforation. A promptly switching to alternative drugs when intestinal tract is involved should be considered in order to prevent surgery.

KEYWORDS: Churg-Strauss System, Allergic Granulomatous Angiitis, Intestinal perforation, Steroids.

CASE REPORT

A 69 year old man with a 5 year history of severe Churg Strauss Syndrome (CSS) was admitted to emergency department because of abdominal pain, respiratory and cardiac insufficiency and a new worsening of oral aphtae. He referred a 5 day history of continuous diarrhea and an episode of rectal bleeding 2 days before. It was considered as a relapse of CSS. Abdominal x-ray was normal and a steroid pulse was given (900 mg of methyl-prednisolone). 2 days after the admittance, the patient complained of an acute diffuse abdominal pain and TC scan showed free-air in sub-diaphragmatic region (Fig. 2). We performed an emergency laparotomy finding multiples colonic perforations localized in the right and transverse colon. Resection of right and transverse colon with an end type ileostomy was performed. Multiple ulcers, extravasal granulomas and mucosal pseudo-polyps were found in the specimen (fig. 2). Bowel function through ileostomy took place on the second post-operative day and oral feeding was started on the fifth day. Laparotomy wound infection occurred on the fourth post operative day. It was successfully treated with antibiotics and Vacuum Assisted Closure (V.A.C.) therapy. One month later the patient was discharged in good general conditions and with normal ileostomy function.



Fig. 1: Transverse CT images showing free air in abdomen

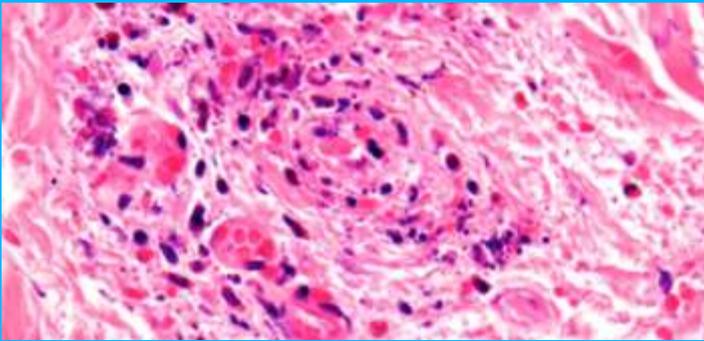


Fig. 2: Extravasal granuloma founded in the specimen

DISCUSSION

CSS is a rare vasculitis that affects small to medium sized vessels associated with asthma and eosinophilia [1]. In 1990 the American College of Rheumatology proposed six criteria for classifying CSS with four being necessary to be diagnosed with a sensitivity of 85% and a specificity of 99.7% [2]. These criteria are asthma, eosinophilia greater than 10% on differential white blood cell count, mononeuropathy (including multiplex) or polyneuropathy, migratory or transient pulmonary infiltrates detected radiographically, paranasal sinus abnormality and biopsy containing a blood vessel with extravascular eosinophils [1].

Gastrointestinal system is involved in almost 31% of patients and it is one of the poor-prognostic factors best known [3]. Small bowel appears to be more frequently involved (Table 1) [4-6]. Clinical manifestations of bowel localization usually consist on abdominal pain and gastrointestinal disorders [7]. Ulcerations are often found in endoscopy and they are considered as precursors of perforation. Review of previous publications and distribution of gastrointestinal tract involvement are reported in table 1. Up to now 18 cases of large bowel involvement are reported, most of them by Japanese authors [5].

Corticosteroids are the cornerstone of initial treatment of CSS [7]. Massive steroid therapy is often used to control disease relapsing, as reported in table 1, but corticosteroids itself enhances fibrotic changes of the intimal tissue, which could consequently cause intestinal perforation due to vascular obstruction [..]. So it can create an environment predisposition to the perforation and mask its initial symptoms. A resection of necrotic intestinal tract is necessary when perforation is confirmed.

Recently alternative immunosuppressive therapies are used to treat patients with poor prognostic factors, or predisposed to relapsing [3, 7]. These drugs and their indications are reported in table 2.

Switching to immunosuppressors should be promptly considered when critical gastrointestinal involvement is showed by symptoms or endoscopy in order to reduce the evolution to perforation, and consequently the number of surgical resection. Moreover during CSS relapsing or when a poor-prognostic factor is already assessed like in our case. Further studies, recruiting a bigger number of patients with

poor-prognostic factors, are needed to confirm the effective possibility of the therapeutic switch to avoid emergency surgery.

Table 1 Review of literature of CSS and bowel involvement

Total of Cases reported (n)	61	Large Bowel involvement	In 29,11 % of cases
Gastrointestinal Involvement	32,6 % of all CSS affected	Small Bowel involvement	In 53,5 % of cases
Mortality	11,9 %	Gastro-duodenal involvement	In 16,6 % of cases
Most frequent symptoms:	Bloody Diarrhea + Abdominal distension	Pancreatitis and Cholecystitis	In 6,3 % of cases

Table 2 Review of recently purposed immuno-modulatory therapy

Drugs	Indications	Duration	Results	Poor-prognostic factor	
intravenous prednisone pulse followed by oral Prednisone	First line therapy	One year	Remission in 93% of patients	none	
Oral Azathioprine	Relapsing in first year	At least 6 months	Good chronic control		
6 Cyclophosphamide pulse every 2 week, then monthly		1+3 months	Best with lower cumulative dosage		
intravenous prednisone pulse plus oral Prednisone plus Cyclophosphamide every 2 weeks then monthly	Relapse control	1+3 months	Remission in 80% of patients	Almost one between: elevated creatinine levels, proteinuria, gastrointestinal/cardiomypathy/central nervous system involvement.	
6 Cyclophosphamide pulse every 2 weeks then monthly (2 months) followed by Azathioprine		One year	Good control with less toxic effects		
Cyclosporine then Azathioprine or ASA	Steroid resistant	3 months	Reduces colectomy rates in the short term.		
Omalizumab	inadequate control persistent allergic asthma	6 months	steroid-sparing agent		

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