up to 50% 5-year survival has been reported.\textsuperscript{3,14} Thus involvement of the anterior abdominal wall does not necessarily mean that these lesions are non-resectable. They are potentially curable lesions in spite of their size and wide excision of all involved structures is recommended where feasible.

References


A case of Churg–Strauss vasculitis complicated by small bowel necrosis

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Summary: A case of Churg–Strauss syndrome causing mesenteric intestinal ischaemia and small bowel necrosis is described in a 29-year-old man. Despite conservative management, the patient’s condition deteriorated and he underwent five laparotomies. Small and medium-sized arteries within the mesentery and lymph nodes showed necrotizing vasculitis. Currently he is doing well on oral nutrition and medical management.

Introduction

Acute intestinal ischaemia is a surgical emergency with life-threatening features. Mesenteric vasculitis is a rare cause of intestinal ischaemia accounting for 2% of cases.\textsuperscript{1} Rheumatoid arthritis, scleroderma, systemic lupus erythematosus, giant cell arteritis, Wegener’s granulomatosis and Churg–Strauss syndrome are systemic diseases which may rarely cause intestinal ischaemia and infarction. We describe a patient with Churg–Strauss syndrome who survived after multiple resections for small bowel necrosis.

Case report

A 29-year-old man with a 5-year history of bronchial asthma and allergic rhinitis was admitted to
Westminster Hospital in June 1992 because of retrosternal pain radiating to the left arm, and epigastric pain associated with nausea. There was no family history of asthma or other atopic diseases.

Physical examination on admission revealed a mild pyrexia of 37.5°C and wheezes throughout both lungs. His abdomen was soft without palpable masses but very tender in the epigastrium. Laboratory examinations showed a white blood cell count of 32.8 × 10⁹/l with a predominant eosinophilia (66.5%) and the erythrocyte sedimentation rate was 86 mm/hour, other haematological parameters were unremarkable. Routine electrolytes were within normal limits, while alkaline phosphatase was elevated at 434 U/l. IgA was 5.3 g/l (normal 0.8–4.0), IgE was 907 U/ml (normal <81), C-reactive protein 68 mg/l (normal <10.0). Anti-nuclear, anti-mitochondrial, anti-smooth muscle, anti-parietal cell, anti-reticulin and anti-neutrophil cytoplasm antibodies were all negative but rheumatoid arthritis (Latex) test was positive (2/1,280). Bone marrow biopsy showed a reactive marrow with an increase in the eosinophilic compartment. Maturation was normal and there was no evidence of a leukaemia process. An initial chest X-ray was unremarkable. However, the electrocardiogram showed possible acute anterior infarction. Abdominal ultrasound was negative but computed tomographic scan showed some small left-sided para-aortic nodes. Upper gastrointestinal endoscopy, and biopsies of stomach and duodenum were all within normal limits, and liver biopsy showed an increased number of eosinophils.

The patient remained on the ward for 20 days, during which he received intravenous fluids, bronchodilators and ranitidine. His chest pain resolved, but the abdominal pain became steadily worse, the white blood cell count raised to 45 × 10⁹/l and C-reactive protein to 462 mg/l (normal <10.0).

At this stage (20 days post-admission). Churg–Strauss syndrome was suspected and the patient received an intravenous bolus dose of 1 g of cyclophosphamide with 1 g of methylprednisolone. The same dose of corticosteroid was repeated on the two following days. As his abdominal pain had not resolved, a laparoscopy was performed under general anaesthesia which showed diffuse peritonitis with extensive areas of small bowel necrosis. Laparotomy confirmed the above findings and a good major vessel blood supply to the small bowel was noted. It was clear that resection of a major portion, certainly more than 70%, of the small bowel would be necessary. It was therefore decided, since arteries and veins right up to the small bowel itself were functioning well, that the entire small bowel would be returned to the peritoneal cavity in order to see if these areas of questionable viability would improve with steroid treatment. A policy of repeated laparotomies was formulated, the timing of which would be determined by the patient’s progress, with particular reference to sepsis. Biopsies were taken from the small bowel and mesenteric lymph nodes.

The second laparotomy 2 days later required a conservative small bowel resection of 30 cm of jejunum and 30 cm of ileum which was irrevocably gangrenous. In addition the residual small bowel had numerous patches of necrosis some of which were locally excised.

Small intestinal biopsy showed focal mucosal ulceration with vasculitis affecting medium and small arteries in the mesentry and submucosa; adjacent veins were not involved (Figure 1). Within the walls and in the adjacent connective tissue, there was a dense, eosinophilic infiltrate extending into the muscularis propria and serosa. The lymph nodes also showed evidence of vasculitis and there was a prominent sinus histiocytosis and a mild oleogranulomatous reaction.

The third laparotomy was performed on day 3 and a further resection was required leaving 10 cm of jejunum and 80 cm of ileum. The fourth laparotomy was carried out on day 6 as perforation was suspected clinically. This proved not to be the case but four small ischaemic plaques were excised.

Specimens showed similar features to the original biopsy, except that they were more florid; the ulceration was more severe and the vessels more extensively necrotic. Infectious agents had been sought by special staining techniques; there was no evidence of helminthic infections, Treponema pallidum, Gram-positive or -negative organisms, cytomegalovirus, hepatitis surface antigen or any other pathogen.

In the time between laparotomies, the patient remained in the intensive care unit receiving total parenteral nutrition and antibiotics. Ten days after he returned to the ward he underwent a fifth

![Figure 1](http://pmj.bmj.com/)

**Figure 1** Mucosal ulcer with underlying inflamed and thrombosed artery. Note adjacent uninvolved vein (H & E, × 25).
Discussion

Intestinal vasculitis is an uncommon cause of small bowel ischaemia and necrosis. Rheumatoid arthritis, scleroderma, polyarteritis nodosa, systemic lupus erythematosus, giant-cell arteritis, Wegener’s granulomatosis and Churg–Strauss syndrome are a group of disorders which share inflammatory and necrotic features of blood vessels, and may involve the mesenteric circulation.

Allergic granulomatosis and angiitis (or Churg–Strauss syndrome) is a rare disease of unknown cause. The syndrome is characterized by hypereosinophilia of both blood and tissues, and a systemic vasculitis in patients with allergic asthma or rhinitis. Although the classical lesion described by Churg and Strauss is the ‘allergic granuloma’, its absence does not exclude the diagnosis. The complete description of the clinical features and course of the disease has been reported by Lanham and co-workers. The differential diagnosis of vasculitis is very difficult and many patients have been described as having vasculitis of unknown cause. In addition many authors feel there is a close relationship between Churg–Strauss syndrome and many other granulomatous, vasculitic and eosinophilic disorders, and Churg–Strauss syndrome is considered as a point of overlap between these three disease spectra. The American College of Rheumatology developed criteria for the classification of Churg–Strauss syndrome. However, these criteria will change when the aetiology and pathogenesis of these vasculitic disorders are better understood.

In the English language literature, the gastrointestinal manifestations of Churg–Strauss syndrome include eosinophilic enteritis, but in the majority mucosa ulceration was not present. On the other hand, Japanese cases are characterized by multiple ulcers which were considered to be caused by ischaemia secondary to vasculitis, but most of them had not been diagnosed until laparotomy or autopsy. In a large series of 165 patients, 52 (31%) had abdominal signs and symptoms. The most severe episodes of abdominal pain were due to peritonitis (nine cases), duodenal ulcer (three cases), intestinal infarction (one case) or unexplained pain (one case), and 11 patients presented with gastrointestinal haemorrhage. However, these patients had systemic vasculitis of the polyarteritis nodosa (PAN) type and Churg–Strauss angiitis as described in Fauci’s classification. The authors did not separate the two conditions because they believe these two forms of angiitis belong to the same group. This theory has also been supported by other workers.

In another series of 106 patients with vasculitis of various causes, 38 patients (36%) had gastrointestinal manifestations. However, intestinal infarction was found in only three out of 13 patients with polyarteritis nodosum, and none with leucocytoclastic vasculitis.

A third series, reported by Camilleri et al., 18 out of 65 patients with systemic vasculitis had gastrointestinal involvement including abdominal pain (85%), diarrhoea (50%), and gut haemorrhage (44%). This was confirmed histologically in 11 patients by finding non-specific inflammation (nine patients) and ulceration (two patients).

Our patient had the typical features of Churg–Strauss syndrome, including allergic rhinitis, history of bronchial asthma, eosinophilia, systemic vasculitis and raised concentrations of IgE. It is interesting that all the IgE values in one series were elevated. This elevation supports the theory that an allergic mechanism is always present in this syndrome.

Should a patient present with abdominal signs due to vasculitis, initial treatment with cyclophosphamide could possibly prevent the mesenteric ischaemia, as mesenteric vasculitis is associated with a poor prognosis. Bacon and co-workers reported a decrease in both mortality and relapse rate in more than 100 cases of systemic necrotizing vasculitis using intermittent pulsed intravenous cyclophosphamide therapy. Intravenous combination of cyclophosphamide and methylprednisolone has also been proved very effective in patients with systemic vasculitis. In a prospective randomized trial of 71 patients the addition of cyclophosphamide to a regime of corticosteroids and plasma exchange has shown a reduction in the incidence of relapses, in patients with polyarteritis nodosum and Churg–Strauss vasculitis, without affecting the 10-year survival rate. However, it is felt this should be confirmed with a multicentre controlled prospective trial.

Our patient received his first course of cyclophosphamide and prednisolone 20 days after the onset of his abdominal symptoms, and a second course after his last laparotomy. From a surgical point of view this patient retained the maximum length of small bowel that was conceivable possible. The policy of repeated laparotomy, though tedious, time consuming, expensive and hazardous from a sepsis point of view, nevertheless ensured...
that any bowel capable of recovery was preserved. The problem with small bowel vasculitis is that the ischaemic process does not always affect the full thickness of the bowel. The mucosa may be lost with survival of the muscle and the lamina propria. In this situation mucosa can regenerate and the bowel be preserved. We are convinced that this policy was responsible for the small bowel preservation. Small bowel ischaemia or necrosis due to Churg–Strauss syndrome is a rare condition that requires prompt medical treatment and surgical intervention, if necessary.

References

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