

# Intestinal Involvement in Wegener's Granulomatosis

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## Abstract

Wegener's granulomatosis is a necrotizing vasculitis that may affect any viscera. Gastrointestinal involvement is rather uncommon. We present a case of Wegener's granulomatosis with multiple small bowel ulcers and a single ileal perforation. Histological examination of the surgically resected specimen demonstrated evidence for necrotizing vasculitis. Gross and histological features of the gastrointestinal disease in Wegener's granulomatosis are discussed.

## Key words

Wegener's granulomatosis - small intestine - intestinal perforation

## Introduction

Wegener's granulomatosis is a small vessel vasculitis that predominantly affects the upper and lower respiratory tract and the kidneys. Gastrointestinal involvement is rather uncommon and is presented with mild to severe life-threatening complications (1). To our knowledge, only a few cases with intestinal perforation have been reported in the English language literature (1-3).

We present a case of Wegener's granulomatosis with multiple small intestinal ulcers and perforation.

## Case report

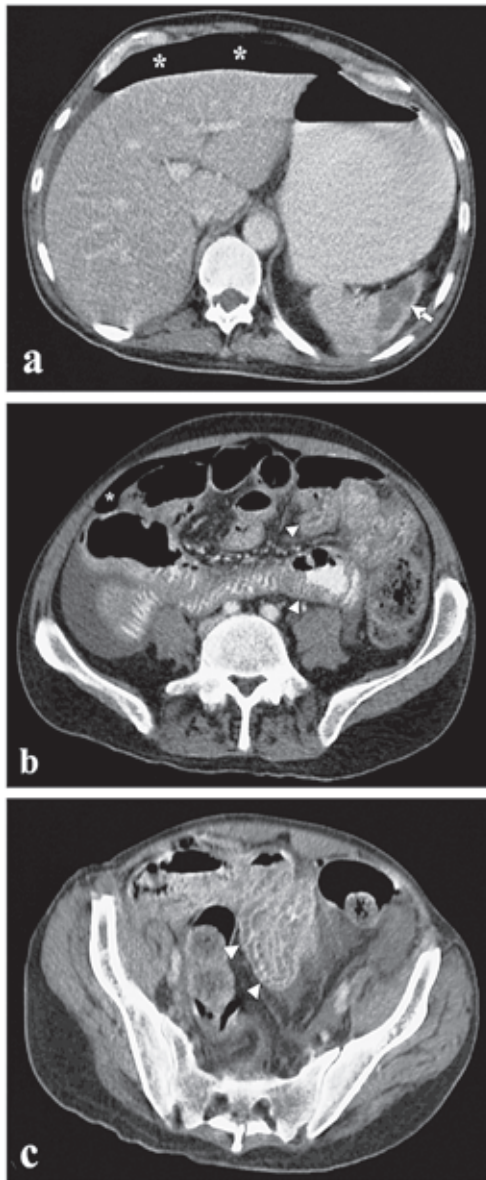
A 44 year-old man had a one month history of Wegener's granulomatosis with multiple nasal nodules, pulmonary cavities and serum ANCA positivity (1.53 mg/dl; N: 0-0.8mg/dl). The patient was admitted to the hospital because of abdominal pain. On admission abdominal examination

revealed diffuse rebound, tenderness and diminished bowel sounds. Complete blood count showed leucocytosis (18000/mm<sup>3</sup>), normal hemoglobin level and platelet count. Other laboratory tests were within normal limits. Contrast enhanced computerized tomography scan showed intraabdominal free air and fluid consistent with gastrointestinal tract perforation. Small bowel wall thickening was identified and chronic splenic infarction was detected (Fig.1a,b,c). The patient underwent an emergency operation. During laparotomy, a perforation area in the ileum was observed and segmental ileal resection was performed.

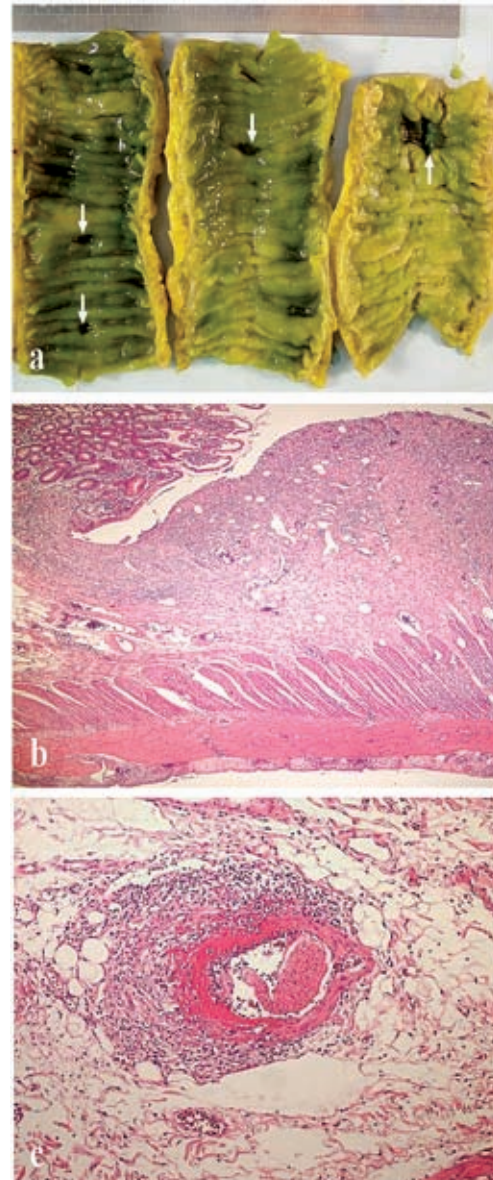
The surgically resected small intestine measured 110 cm in length. The mucosal surface of the small intestine showed multiple transversely oriented shallow ulcers (Fig.2a). One of the ulcerations was perforated. Histological examination of the resected intestinal specimen revealed ulcerations and necrotizing granulomatous vasculitis of the small to medium sized vessels most notably involving the submucosal blood vessels. The vasculitis appeared as transmural and granulomatous inflammation accompanied by fibrinoid necrosis (Fig.2b,c). Granulomas were not observed throughout the intestinal wall. Suppurative lymphadenitis was found in the mesenteric lymph nodes. Immunohistochemical study for cytomegalovirus antigen was negative and acid-fast stain was also negative in tissue sections. The performed skin biopsy revealed vasculitis. The patient has been under control with oral systemic steroid therapy for 4 months.

## Discussion

Wegener's granulomatosis is characterized by necrotizing vasculitis and granulomatous inflammation (1). The disease has a predilection to involve upper and lower respiratory tract and the kidneys. In the clinical course of the illness most patients show multisystemic disease (4). However, intestinal involvement is uncommon and usually detected in autopsy studies (5). Classical descriptions of intestinal pathology of Wegener's granulomatosis have been based on the study of surgically resected specimens.



**Fig.1** Intravenous contrast enhanced CT images (a, b, c): free intraabdominal air (asteriks) and fluid secondary to intestinal perforation. Accompanying chronic splenic infarction (arrow). Prominent wall thickening of small bowel with mucosal and serosal enhancement separated by low attenuation zone representing mural edema (arrow heads).



**Fig.2** a. Photograph of the small bowel showing multiple transverse ulcers (arrows). b. Histologic section of the small bowel demonstrating ulcers (HE, x40) and (c) necrotizing vasculitis in the submucosal layer (HE, x200).

Pathological findings include multiple small ulcerations, and intestinal perforation. Histological examination usually shows marked mixed inflammatory infiltrate associated with necrotizing and granulomatous vasculitis of small-medium sized vessels. The small bowel is the most common site (1,6).

In the literature, there are conflicting reports about intestinal involvement in Wegener's granulomatosis. It is suggested that the use of corticosteroid therapy may be an aetiological factor for the development of intestinal manifestations (7). In some reported cases, the main problem is to determine the causative agent for intestinal involve-

ment. Some of the authors suggested that in order to establish the diagnosis of intestinal Wegener's granulomatosis there should be histologic evidence of vasculitis (8). Our findings in this case are in line with the reported cases that show vasculitic changes (2,8).

Clinically, gastrointestinal Wegener's granulomatosis mimics inflammatory and infectious bowel diseases (9). In clinical practice the role of endoscopic mucosal biopsies in the diagnosis of Crohn's disease and tuberculosis is sometimes limited and may only demonstrate nonspecific inflammatory changes (10). Presumptive diagnosis based on clinical and gross features remains important.

CT findings of bowel involvement are nonspecific for the differentiation of Wegener granulomatosis, unlike other types of small-vessel vasculitides. Diffuse or multifocal bowel wall thickening, abnormal enhancement pattern of bowel wall, dilatation of bowel segments and mesenteric vessel engorgement and ascites can be included among those nonspecific findings. As in our case, the presence of perforation can be effectively demonstrated in such cases (11,12).

The ulcer formation of Wegener's granulomatosis has been considered to be caused by vasculitis that impairs the blood supply. Descriptions of the ulcer morphology in the diseases with intestinal involvement were based on the study of surgically resected specimens (8,9). In our case, the pathologic findings in the intestine were multiple, small, shallow and clear ulcerations with less prominent exudate. In contrast to those of typhoid fever, these ulcers were transversely oriented and localized especially between the villous structures of the small intestine. But they were similar to those of intestinal tuberculosis (13). Microscopic findings were more characteristic and more helpful than in other ulcerative intestinal diseases. The surrounding mucosa adjacent to the ulcerations did not show fibrotic and ischemic changes. Tubercular ulcers are also transverse but they are accompanied by fibrosis extending through the wall which may be multiple or single (13). In Crohn's disease, intestinal ulcers are longitudinal or serpiginous and in a segmental distribution (14). The gross and microscopic features of the ulcers in Wegener's granulomatosis, which were shallow and transversely oriented, can be helpful in making the distinction between Crohn's disease and tuberculosis.

Histologically, another problem is to differentiate Wegener's granulomatosis from other forms of vasculitis such as Churg-Strauss syndrome and polyarteritis nodosa, which show histologic changes similar to Wegener's granulomatosis. The interpretation of both clinical and morphological data should be appropriate (15).

In conclusion, the morphologic features of the ulcers and intestinal involvement in Wegener's granulomatosis may be helpful in distinguishing this entity from different entities such as Crohn's disease and tuberculosis, but not from Churg-Strauss syndrome and polyarteritis nodosa. It is essential to find the histological evidence of vasculitis in

intestinal specimens to support the diagnosis of Wegener's granulomatosis.

## References

1. Storesund B, Gran JT, Koldingsnes W. Severe intestinal involvement in Wegener's granulomatosis: report of two cases and review of the literature. *Br J Rheumatol* 1998; 37: 387-390.
2. Geraghty J, Mackay IR, Smith DC. Intestinal perforation in Wegener's granulomatosis. *Gut* 1986; 27: 450-451.
3. Haworth SJ, Pusey CD. Severe intestinal involvement in Wegener's granulomatosis. *Gut* 1984; 25: 1296-1300.
4. Hoffman GS, Kerr GS, Leavitt RY, et al. Wegener granulomatosis: an analysis of 158 patients. *Ann Intern Med* 1992; 116: 488-498.
5. Walton EW. Giant-cell granuloma of the respiratory tract (Wegener's granulomatosis). *Br Med J* 1958; 2: 265-270.
6. Chow FY, Hooke D, Kerr PG. Severe intestinal involvement in Wegener's granulomatosis. *J Gastroenterol Hepatol* 2003; 18: 749-750.
7. Akça T, Çolak T, Çadlıkülekçi M, Öcal K, Aydın S. Intestinal perforation in Wegener's granulomatosis: a case report. *Ulus Travma Derg* 2005; 11: 348-351.
8. Tokuda M, Kurata N, Daikuhara H, et al. Small intestinal perforation in Wegener's granulomatosis. *J Rheumatol* 1989; 16: 547-549.
9. Veinot JP, Logan CA, Thomas MJ. Wegener's granulomatosis arteritis causing small bowel infarction. *Pathology* 2003; 35: 268-269.
10. Pulimood AB, Peter S, Ramakrishna BS, et al. Segmental colonoscopic biopsies in the differentiation of ileocolic tuberculosis from Crohn's disease. *J Gastroenterol Hepatol* 2005; 20: 688-696.
11. Frauenfelder T, Wildermuth S, Marincek B, Boehm T. Nontraumatic emergent abdominal vascular conditions: advantages of multi-detector row CT and three-dimensional imaging. *Radiographics* 2004; 24: 481-496.
12. Pickhardt PJ, Curran VW. Fulminant enterocolitis in Wegener's granulomatosis: CT findings with pathologic correlation. *AJR Am J Roentgenol* 2001; 177: 1335-1337.
13. Patel N, Amarapurkar D, Agal S, et al. Gastrointestinal luminal tuberculosis: establishing the diagnosis. *J Gastroenterol Hepatol* 2004; 19: 1240-1246.
14. Sharma MP, Bhatia V. Abdominal tuberculosis. *Indian J Med Res* 2004; 120: 305-315.
15. Kim YB, Choi SW, Park IS, Han JY, Hur YS, Chu YC. Churg-Strauss syndrome with perforating ulcers of the colon. *J Korean Med Sci* 2000; 15: 585-588.