Rare disease

Giant cell phlebitis: a potentially lethal clinical entity

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Summary

An 83-year-old woman presented to us with a 4-week history of general malaise, subjective fever and lower abdominal pain. Despite the intravenous infusion of antibiotics, her blood results and physical condition worsened, resulting in her sudden death. Autopsy study revealed that the medium-sized veins of the mesentery were infiltrated by eosinophil granulocytes, lymphocytes, macrophages and multinucleated giant cells; however, the arteries were not involved. Microscopically, venous giant cell infiltration was observed in the gastrointestinal tract, bladder, retroperitoneal tissues and myocardium. The final diagnosis was giant cell phlebitis, a rare disease of unknown aetiology. This case demonstrates for the first time that giant cell phlebitis involving extra-abdominal organs, including hearts, can cause serious morbidity.

BACKGROUND

Giant cell phlebitis is a localised vasculitis of the bowel wall and mesentery; patients usually present with symptoms of subacute-to-acute visceral ischaemia. The clinical and radiological features are non-specific, and exploratory surgery is usually required for histological diagnosis; bowel resection is often both diagnostic and therapeutic.

Prior to this report, giant cell phlebitis was thought to usually follow a benign course, and was not known to involve extra-abdominal organs.

CASE PRESENTATION

An 83-year-old woman presented to us with a 4-week history of general malaise, subjective fever and lower abdominal pain. These symptoms were preceded by over 2 months of appetite loss and epigastralgia. Her clinical history was otherwise unremarkable.

INVESTIGATIONS

Physical examination revealed a soft and diffusely tender abdomen, with normal bowel sounds. The leucocyte count was 12 060 cells/ μ l and the C-reactive protein (CRP) level was 17.65 mg/dl. The remaining laboratory

data were normal, with a negative antineutrophil cytoplasmic antibody test. Abdominal CT showed no significant findings (figure 1A).

DIFFERENTIAL DIAGNOSIS

The patient's 4-week history of fever and abdominal pain suggested inflammatory bowel disease, colon cancer or gynaecological diseases. On the other hand, the high levels of CRP level strongly suggested inflammation due to bacterial infection.

TREATMENT

Treatment with ceftriaxone, a broad-spectrum antibiotic, and intravenous fluids was initiated. On day 7, her abdomen became distended and diffusely tender with involuntary guarding. Despite antibiotic treatment, her leucocyte count had increased to 22 410 cells/ μ l, and her CRP to 20.63 mg/dl. Her fever persisted, and the worsening inflammation prompted us to repeat a contrast-enhanced abdominal CT, showing that the bowel walls were now oedematous and thickened (figure 1B). Colonoscopy indicated that the sigmoid colon and rectum were narrowed and oedematous; endoscopic studies

Figure 1 Abdominal CT. (A) Normal bowel wall at admission. (B) On day 7, the bowel wall became thickened and oedematous (white arrow).

Figure 2 Multinucleated giant cells were observed around the veins (Ve) of the bowel. The arteries (Ar) were not involved.

revealed highly restricted mobility of the stomach and colon. These findings suggested ischaemic colitis as the aetiology for the patient's critical condition. Despite the addition of intravenous immunoglobulin to her antibiotic treatment, her blood results and physical condition remained unchanged.

OUTCOME AND FOLLOW-UP

On day 15, the patient died suddenly.

At autopsy, the mesentery was found to be shrunken and hard. The medium-sized veins of the mesentery were infiltrated by eosinophil granulocytes, lymphocytes, macrophages and multinucleated giant cells; however, the arteries were not involved (figure 2). The gastrointestinal tract was severely immobilised because of the hardened mesentery, but the bowel wall did not show macroscopic evidence of thickening or infarction. Microscopically, venous giant cell infiltration was observed in the gastrointestinal tract, bladder, retroperitoneal tissues and myocardium (figure 3). The myocardial inflammation was considered likely to have caused a fatal arrhythmia, resulting in sudden death.

DISCUSSION

Giant cell phlebitis is characterised by localised vasculitis of the bowel wall and mesentery, without arterial

Figure 3 Inflammatory cell infiltration, including that of multinucleated giant cells, was observed in the myocardium.

involvement or evidence of systemic vasculitis.¹ Its pathogenesis remains unknown. The clinical, radiological and endoscopic findings are often found to be in conflict. The salient feature of the disease is subacute-to-acute visceral ischaemia manifesting as abdominal pain, haematochezia and bloody diarrhoea. Patients often approach or are referred to gastroenterologists, and a work-up for inflammatory bowel disease is usually initiated. Although the differential diagnosis includes mesenteric vasculitis associated with systemic lupus erythematosus, Behçet's disease and Churg-Strauss syndrome, these conditions primarily involve the arteries, not the veins. Abdominal CT findings of thickened and oedematous bowel walls have been reported in giant cell phlebitis, which unfortunately cannot be considered of diagnostic value.² Histological diagnosis of the disease requires an assessment of submucosal vessels, which are difficult to obtain using endoscopic biopsy.³ To our knowledge, fewer than 50 cases of giant cell phlebitis cases have been reported, all of which were refractory to medical management and required surgical exploration. These cases involved artery-sparing vasculitis limited to the gastrointestinal tract, with obvious phlebitis in the resected bowel. Surgical resection of the affected bowel is usually both diagnostic and curative, without recurrences.⁴ In the present case, the most severe inflammation was observed in the mesentery rather than the bowel itself, suggesting that the disease had already disseminated to multiple tissues. Because the disease often has a non-specific presentation, preoperative diagnosis is quite difficult. Although the disease usually runs a benign course, this case demonstrates shows for the first time that the giant cell phlebitis can involve extra-abdominal organs, including the heart, and can cause serious morbidity. Clinicians should, therefore, be aware of this rare clinicopathological entity.

Learning points

- ▶ Giant cell phlebitis is characterised by localised vasculitis of the bowel wall and mesentery, without arterial involvement or evidence of systemic vasculitis.
- ▶ Surgical resection of the affected bowel is usually both diagnostic and curative.
- ▶ Giant cell phlebitis can involve extra-abdominal organs, including the heart, and can cause serious morbidity.

Competing interests None.

Patient consent Obtained.

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