

Case report

Double pylorus in a patient with Behçet's syndrome

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ABSTRACT

We report a patient with Behçet's syndrome who presented with upper gastrointestinal haemorrhage. Gastroduodenoscopy showed a gastroduodenal fistula which caused the appearance of double pylorus in the antrum. The possibility of peptic ulcer disease related to non-steroidal anti-inflammatory drug use or Behçet's syndrome itself, as the cause of this rare condition in this patient is discussed.

Introduction

Behçet's syndrome (BS) is a systemic vasculitis characterised by oral ulcers, genital ulcers, papulopustular lesions, erythema nodosa, positive pathergy reaction, eye involvement, arterial aneurysms, deep and superficial vein thrombosis, neurologic involvement and gastrointestinal involvement. Patients with gastrointestinal involvement usually present with abdominal pain and diarrhea and less frequently with bleeding and perforations. The most commonly involved areas are the ileocolonic region and the colon. Endoscopy usually shows single large ulcers. It may often be clinically, endoscopically and histologically impossible to differentiate it from Crohn's disease (1). The prevalence shows wide geographic variation. The frequency of gastrointestinal involvement is reported as high as 25% in series from Japan and Korea, whereas the frequency is around 1–3% in series from countries around the Mediterranean basin (2). This wide variation could be associated with genetic or environmental factors as well as differences in the subspecialties these reports come from, and whether other causes such as non-steroidal anti-inflammatory drug (NSAID) use that can mimic gastrointestinal involvement are excluded.

Case report

A 45-year-old woman with BS was referred to the endoscopy unit for up-

per gastrointestinal system endoscopy. She was diagnosed with BS 7 years ago with oral ulcers, papulopustular lesions, erythema nodosa, a genital ulcer and pathergy positivity. She was using colchicine 3 times a day for her mucocutaneous symptoms. She had a haematemesis 2 weeks before she presented to our unit. She was receiving tenoxicam for back pain at that time. Endoscopy that was performed in another hospital showed a duodenal and antral ulcer without any signs of recent bleeding. She was prescribed a proton pump inhibitor, and used it for 2 weeks. When she presented to our clinic we performed an upper gastrointestinal endoscopy for checking whether the ulcer had healed. During endoscopy we detected two pyloric channels, one of them was actually a fistula between the antrum and the duodenal bulb (Fig. 1). The antral and duodenal opening of the channel were ulcerated. Figure 2 shows the radiologic image which also demonstrates the fistula between the antrum and the duodenal bulb. Histologic examination of biopsy samples from the ulcer margin showed active erosive gastritis with mild signs of chronicity, foveolar hyperplasia and foveolar epithelial hyperplasia, and regenerative changes in the superficial epithelium. *Helicobacter pylori* was positive in the samples obtained with endoscopic mucosal biopsy from antrum. Her colonoscopy, including the terminal ileum was normal and she had no other lesions suggesting gastrointestinal involvement of BS. Her acute phase reactants were not elevated and her blood count and biochemistry results were normal. She continued her proton pump inhibitor and *Helicobacter pylori* eradication treatment was given. Specific treatment for gastrointestinal involvement of BS such as 5-ASA preparations or azathioprine was not given. A control upper gastrointestinal endoscopy was performed 6 months later and showed

Competing interests: none declared.

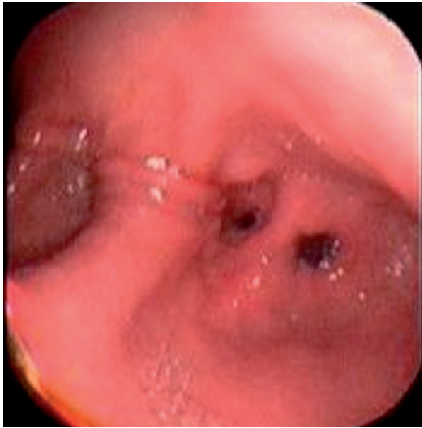


Fig. 1. Endoscopic image showing two openings at the antrum.



Fig. 3. Control endoscopy showing deformed pyloric channel without fistula 9.

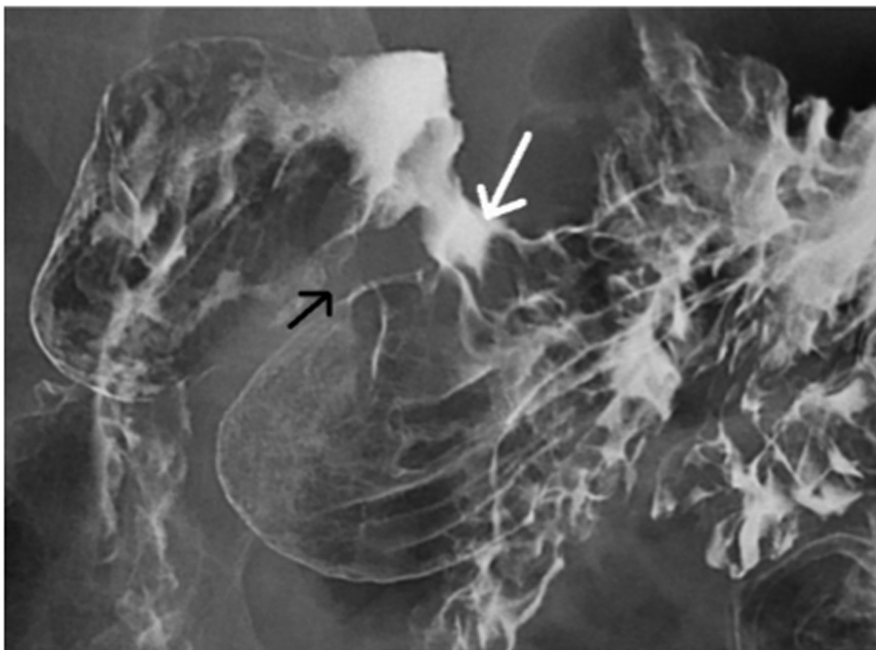


Fig. 2. Upper gastrointestinal barium x-ray showing the gastroduodenal fistula (white arrow) and the pyloric channel (black arrow).

that the ulcer had healed, but the double pylorus appearance persisted. On her final control upper gastrointestinal endoscopy, 1 year after the first one, the fistula had disappeared and a single deformed pylorus with a large opening was seen (Fig. 3).

Discussion

This condition which is generally called "double pylorus" is an uncommon finding. Double pylorus indicates 2 different conditions: the first one is the congenital form which is a very rare condition that has been reported in less than 10 cases (3), and the second one is the acquired form. Acquired

double pylorus is in fact a gastroduodenal fistula presenting usually between the lesser curvature of the gastric antrum and the superior wall of the duodenal bulb. The incidence of this rare condition was 0.02% in the largest series (4). Acquired double pylorus is usually a complication of the peptic ulcer disease. NSAID use has also been reported in some of these cases. In the series by Hu *et al.*, in approximately 27% of the cases, double pylorus transformed into a single deformed pyloric channel (4).

BS is a vasculitis which can also involve the gastrointestinal system. Gastrointestinal involvement usually

causes ulcers in the ileocecal region and colon which may result in bleeding or perforations, somewhat more frequently than that in Crohn's disease. 5-ASA preparations and immunosuppressives such as azathioprine are the mainstay of treatment. Biologics can be used in resistant cases. Gastrointestinal fistulas such as tracheoesophageal, enteroenteric or cecocolic fistulas (5, 6) were rarely reported in BS. Also a BS patient with double antral ulcers forming a mucosal bridge, that can be accepted as a kind of fistula (7) was reported. However, as far as we know, no gastroduodenal fistulas have been reported up to now.

In this patient it is difficult to decide whether the double pylorus is the result of the peptic ulcer disease related to NSAID use or a manifestation of gastrointestinal involvement of BS. The non-specific histologic findings and the absence of acute phase response do not exclude the diagnosis of gastrointestinal involvement of BS because histologic findings may not distinguish BS from other conditions (1) and acute phase response may be normal in BS patients with active gastrointestinal involvement (8). It may be argued that peptic ulcer disease and NSAID use is the cause since her ulcers healed with proton pump inhibitor therapy and *H. pylori* eradication, without immunosuppressive use. Moreover, there were no other manifestations of gastrointestinal involvement of BS in the more commonly involved parts of the gastrointestinal tract such as the ileocolonic region and colon. However, it is possible that the pathergy phenomenon, an enhanced inflammatory response to a traumatic insult or inflammatory stimuli which is quite specific to BS (9), may have played a role in the development of this rare endoscopic sign in this patient.

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