Isolated Gastroduodenal Crohn's Disease

Matthew Sullivan DO
Lehigh Valley Health Network, Matthew.Sullivan@lvhn.org

Patrick Hickey DO
Lehigh Valley Health Network, Patrick.Hickey@lvhn.org

Saba Ahmad MD
Lehigh Valley Health Network, Saba.Ahmad@lvhn.org

Jeffrey Brodsky MD
Lehigh Valley Health Network, Jeffrey_T.Brodsky@lvhn.org

Hiral N. Shah MD
Lehigh Valley Health Network, hiral_n.shah@lvhn.org

See next page for additional authors

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Authors
Matthew Sullivan DO, Patrick Hickey DO, Saba Ahmad MD, Jeffrey Brodsky MD, Hiral N. Shah MD, and Ronald Bross MD

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Matthew J. Sullivan DO, Patrick Hickey DO, Saba Ahmad MD, Jeffrey Brodsky MD, Hiral Shah MD, and Ronald Bross MD

Department of Internal Medicine, Lehigh Valley Health Network, Allentown, PA

Background

- Crohn's disease (CD) is a chronic idiopathic inflammatory disease of the gastrointestinal (GI) tract characterized by segmental and transmural involvement which can occur anywhere along the GI tract.[1] However, there is a predilection for the distal small bowel and proximal colon.[2] CD can also affect extra-intestinal locations including the skin, bones, eyes, liver, gallbladder, urinary tract, and vascular system.[3] The mean age at time of presentation is 20 to 30 years.[4]
- The etiology and pathogenesis are not well understood, but current research indicates that CD is likely related to an abnormal immunological response to natural intestinal flora in genetically-susceptible patients. Genetic, epigenetic, and environmental factors are thought to also play a role and cigarette smoking has been identified as an independent risk factor.[2]
- Active CD is characterized by focal inflammation and fistula formation resulting in fibrosis, stricturing, and thickening of the bowel wall which can lead to recurrent obstructions or chronic fistulas. Presenting symptoms depend on the portion of the GI tract which is affected, but include abdominal pain, diarrhea, nausea, vomiting, and weight loss.[3] Gastroduodenal involvement in CD is typically asymptomatic, but when symptoms do occur the most common is epigastric pain, often postprandial, which can lead to weight loss and anorexia.[4]
- The pathologic hallmark of CD is a granulomatous inflammatory response characterized by focal collections of macrophages, epithelioid cells, and multinucleated giant cells.[2] However, a focal nonspecific acute and chronic inflammation is more common in upper GI tract involvement.[5] The most common pathology finding in gastric CD is a Helicobacter pylori-negative focal gastritis.[4]
- Approximately one-third of patients have ileitis or colitis at the time of diagnosis[2], but only 0.5-4% of all patients with CD have clinically significant gastroduodenal disease.[4] The majority of these patients have concomitant distal disease and there are only a few documented case reports of adults with isolated gastric CD, accounting for only 0.07% of all cases of CD.[1] The first case of gastric CD was described in 1949.[5]
- Imaging in gastroduodenal CD can show aphthous ulcers, "cobblestoning," thickened folds, ulcerations, "ram's horn sign," and a "pseudo-Billroth I appearance" of the antrum and proximal duodenum. Endoscopy with biopsy remains the gold standard for diagnosis.[4]
- Initial treatment of gastroduodenal CD includes corticosteroids and acid suppression with proton pump inhibitor. After remission, 6-mercaptopurine and azathioprine are used for maintenance treatment. One third of patients will require surgery due to duodenal obstruction, refractory pain, fistula formation, or hemorrhage.[4]

Case Presentation

A 69-year-old female former smoker without any known past medical or family history of gastrointestinal disease and malignancy presented with intermittent dysphagia to both solids and liquids associated with severe chest pain.

Initial workup included esophagogastroduodenoscopy (EGD) revealing a small sliding hiatal hernia, mild non-erosive gastritis with benign appearing gastric polyps, and an irregular Z-line which was biopsied to rule out short segment Barrett's esophagus. The duodenum was unremarkable. A colonoscopy was also performed revealing only diverticulosis. She was initiated on proton pump inhibitor (PPI) therapy due to the finding of gastritis.

Two years later she presented with complaints of nausea, epigastric pain, and early satiety over the previous three weeks. A CT scan of the abdomen was performed revealing gastric antral wall thickening and inflammatory stranding.

EGD demonstrated erosions and submucosal hemorrhages in the gastric antrum and pyloric channel. There was also severe bulbar duodenitis with exudate and friability. Multiple biopsies were obtained revealing chronic active inflammation and the patient was maintained on PPI therapy.

Her early satiety progressed and continued weight loss lead to a third EGD with endoscopic ultrasound (EUS). This revealed marked erythematous mucosa with an irregular pattern throughout the antrum with areas of pitting and hypertrophy in a "stalactite" fashion. There was also narrowing of the antrum without gastric outlet obstruction. The echoendoscope revealed 2 cm circumferential thickening of the antrum with loss of native architecture. Using fine needle aspiration, multiple biopsies were obtained of the gastric antrum. There was no evidence of perigastric lymphadenopathy or liver pathology visualized. Biopsies again revealed chronic active inflammation and cytology was negative for malignancy.

At this point the patient was referred to an oncologic surgeon due to concern for possible gastric malignancy. The patient eventually underwent partial distal gastrectomy with Roux-en-Y reconstruction. The surgical specimen was sent for pathology which revealed multiple foci of inflammation with submucosal fibrosis and deep non-caseating granulomas consistent with Crohn's disease.

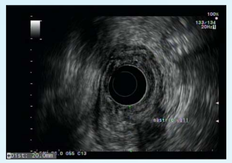

Figure 1. Endoscopic ultrasound revealing circumferential thickening of the gastric wall.

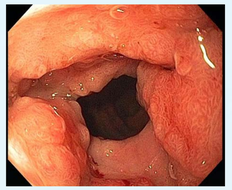

Figure 2. EGD image demonstrating gastritis and erythema of the antrum and marked hypertrophy of the mucosa with narrowing of the pyloric channel.

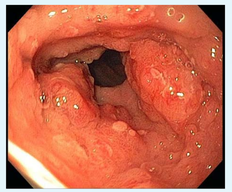

Figure 3. EGD image of the pyloric channel again demonstrating mucosal irregularities and areas of hypertrophy and pitting.

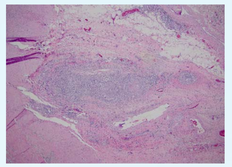

Figure 4. Slide preparation from gastric antrum showing focally necrotizing granulomatous inflammation consistent with Crohn's Disease.

Discussion

This case demonstrates a rare presentation of isolated gastroduodenal Crohn's disease (CD) and the difficulty in making this diagnosis. Nugent and Roy developed criteria for diagnosing gastroduodenal CD which include:

- Noncaseating granulomatous inflammation of the stomach or duodenum and the absence of other systemic granulomatous disorders, or
- Confirmed CD elsewhere and radiographic or endoscopic evidence of CD involvement of the stomach or duodenum.[4]

Significant gastroduodenal involvement occurs in only 0.5-4% of all CD patients and even less have isolated disease.[4] However, due to advancement of diagnostics, non-significant incidence is thought to be much more prevalent.[5]

Examination for lower gastrointestinal tract involvement and malignancy must be ruled out prior to making the diagnosis. Close follow up is also imperative as one-third of patients with gastroduodenal CD will develop distal disease over time.[4]

The symptoms of gastroduodenal CD can easily be confused with peptic ulcer disease (PUD) and this must also be ruled out.[4] Helicobacter pylori testing should be completed as the most common finding is a nonspecific acute and chronic inflammation which is most often H. pylori-negative.[5] Also, ulcerations in CD are typically linear as opposed to the circular ulcers seen with PUD.

Initially our patient did not meet the Nugent and Roy criteria. Under these circumstances anti-Saccharomyces cervisia antibody testing may be beneficial and this was indeed positive in our patient.[3] Despite repeated imaging and endoscopies, our patient did not receive a diagnosis until after surgical resection, demonstrating the complexity in making the diagnosis of isolated gastroduodenal Crohn's disease.

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