Isolated gastroduodenal Crohn disease

Clinical History:

A 20-year-old male presented with recurrent vomiting, low-grade fever and weight loss for the last 6 months. No history of any abdominal pain, NSAID intake, or corrosive intake was present. Fever was low-grade with evening rise and was associated with loss of appetite and significant weight loss of 10 kilogram over a 6 months interval.

Imaging Findings:

Physical examination revealed mild epigastric tenderness and succussion splash. An upper GI endoscopy revealed thickened gastric folds in lower body and pyloric antrum, with thickened duodenal folds in second part of duodenum. Barium study confirmed the thickening of gastric and duodenal folds. Follow through study revealed normal appearing jejunal and ileal loops. CT scan of abdomen demonstrated smooth circumferential gastro-duodenal wall thickening. No significant perigastric lymphadenopathy was evident. Small bowel loops elsewhere were unremarkable and remaining viscera appeared normal. Prior mucosal punch biopsies from the stomach had revealed non-specific inflammation. In view of repeated negative biopsies, a snare-biopsy was performed which demonstrated multiple noncaseating epithelioid granulomas and multinucleated giants cells. The specimen was excessively sampled but no evidence of malignancy was seen. His chest radiograph was normal, however, Mantoux test came positive. Biopsy specimen was subjected to polymerase chain reaction (PCR) for Mycobacterium tuberculosis which was negative ruling out enteric tuberculosis. Serum calcium and ACE levels were normal excluding sarcoidosis. Considering the possibility of Crohn disease he was started on corticosteroids. Within 3 weeks the patient had dramatic relief in symptoms. After 2 months of treatment his steroid dose were tapered. A repeat endoscopy at 6 months revealed significant reduction in the thickness of gastric folds while biopsies showed persistent small mucosal granulomas and mild to moderate lymphoplasmacytic infiltration. No evidence of ileo-colic involvement was seen. Barium enema, colonoscopy with ileoscopy and biopsy were normal. Repeat colonoscopy at 6 months showed normal appearance of colonic and terminal ileal mucosa.

Discussion:

Crohn disease can affect any part of the digestive tract from mouth to anus, most commonly the terminal ileum and proximal colon. Gastroduodenal involvement is highly uncommon (0.5 – 4%) and is relatively more frequent in patients with concomitant distal ileal disease. Isolated gastroduodenal Crohn disease (GCD) is definitely a rare occurrence with only few case reports in the literature. Nugent and Roy criteria for the diagnosis of GCD include either 1) noncaseating granulomatous inflammation of the stomach or duodenum on histopathology, with or without
concomitant Crohn disease in the remaining gastrointestinal tract, and the absence of other systemic granulomatous disorders; or 2) confirmed Crohn disease of the gastrointestinal tract and radiographic or endoscopic findings of diffuse inflammation of the stomach or duodenum consistent with Crohn disease.

While both adult and pediatric population can be affected, it is seen most commonly in the third to fourth decades of life with a male-to-female ratio of 1.2:1. Common presenting symptoms include epigastric pain, vomiting, nausea, anorexia and weight loss. Patient may develop anaemia secondary to chronic blood loss. On endoscopy the gastroduodenal mucosa is usually hyperemic and may show multiple erosions, ulcers or nodular lesions. The lesions predominate in the pyloric antrum and may also involve the body. Proximal extension of the disease is unusual; in contrast duodenum tends to be involved. Advanced stages can lead to stricture or fistula formation. A wide spectrum of radiographic findings may be evident depending upon the disease severity, ranging from mucosal nodularity, ulceration, fold thickening to a typical ‘cobblestone’ mucosa. The earliest radiographic finding includes aphthous ulcers in the gastric antrum or pylorus. These appear as punctate barium collections surrounded by a mound of radiolucent oedema. These may evolve into larger deep ulcers leading to a cobblestone mucosal pattern on upper GI-series. Advanced stages of the disease, owing to scarring, can demonstrate the typical radiographic signs of GCD which include the ram’s horn sign and pseudo-Billroth-I appearance. Ram’s horn sign represents a characteristic funnel-shaped deformity of the pyloric antrum and duodenal bulb. The funnel-shaped stomach has been likened to the sacramental ram’s horn or shofar, used to sound the advent of the Jewish new year. A scarred pylorus and duodenum can form a continuous tubular structure with obliteration of the normal anatomical landmarks. This can mimic the post-operative appearance of Billroth-I partial gastrectomy hence the pseudo-Billroth-I sign. A rigid and narrowed stomach can also present with a linitis plastica like picture. Other entities which can manifest with similar barium findings include peptic ulcer disease, tuberculosis, sarcoidosis, eosinophilic gastroenteritis, Zollinger-Ellison syndrome, gastric carcinoma or lymphoma. Although CT is not diagnostic it can aid in narrowing down the differentials.

A correct diagnosis of GCD is challenging and needs a high degree of clinical suspicion. It can be achieved by combining recognition of clinical features, imaging findings and extensive clinical work-up including endoscopic biopsy. Medical therapy with steroids and immunomodulatory drugs is the first line of management. Balloon dilation and surgery are considered in refractory cases.  

**Differential Diagnosis List:** Isolated gastroduodenal Crohn’s disease

**Final Diagnosis:** Isolated gastroduodenal Crohn’s disease

**References:**


Description: The gastric rugal folds show marked thickening especially in the antrum and lower body of stomach. Duodenal cap is also deformed. Origin:
Description: Circumferential gastro-duodenal wall thickening is well visualised. Perigastric fat planes are preserved. Origin:
Figure 3

Description: Upper GI endoscopy shows oedematous and thickened gastric rugal folds with hyperemic and erythematous mucosa. Origin:
Description: Duodenal folds also appear oedematous and thickened. Origin:
Description: Multiple noncaseating epithelioid granulomas and multinucleated giants cells with marked lymphoplasmacytic infiltration in the gastric mucosa. Origin: