A Rare Case of Isolated Duodenal Crohn’s Disease

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Abstract: Crohn’s disease (CD) is an inflammatory bowel disease that is characterized by segmental and transmural involvement of any portion of the gastrointestinal tract from the mouth to the anus. However, isolated duodenal involvement is rather extremely rare and diagnosis requires comprehensive clinical evaluation. Herein we present the case of a 55-year lady who was referred to the gastroenterology team with a working diagnosis of possible malignant gastric outlet obstruction. The lady had no background medical or surgical history apart from being investigated for iron deficiency anemia years back by having gastroscopy and colonoscopy which were not significant. Her son was known to have Crohn’s disease. She was originally admitted to hospital with a history of upper abdominal pain, early satiety and weight loss. There were no bowel symptoms. Apart from iron deficiency anemia, her blood investigations were normal. The CT abdomen and pelvis showed duodenal wall thickening with a stricture but otherwise normal, including normal bowels. She had repeated gastroscopy, dilatation and sampling of the stricture site. The first two samples showed non-specific duodenitis. The diagnosis of isolated duodenal Crohn’s disease was made based on the samples from the third gastroduodenoscopy procedure which showed non-caseating granuloma. She had normal ileocolonoscopy and biopsies. Her fecal calprotectin was 230 micrograms per milligram (50-200). Based on the above findings she was put on a course of steroids and started on a standard dose of Infliximab infusion following negative surveillance to rule out latent Tuberculosis and to exclude other viruses as per protocol. The patient made marked clinical, endoscopic and histological response thereafter. This case emphasizes the challenge of diagnosing duodenal Crohn’s disease which, as above, might require high index of clinical suspicion and repeated sampling. We expect that this case report adds to the existing literature on this subject.

Keywords: Crohn’s Disease, Duodenal Crohn’s, Isolated Duodenal Crohn’s Disease, Non-caseating Granuloma, Case Report

1. Introduction

Crohn’s disease (CD) is a chronic inflammatory bowel disease. The exact etiology of the disease is not known. It causes transmural inflammation of any part of the gastrointestinal tract from mouth to anus and it can also cause some extraintestinal manifestations that can affect other organs like skin, joints, and eyes. In most cases it affects the terminal ileum or colon and rarely affects the stomach and duodenum. Crohn’s disease of the upper gastrointestinal tract occurs in 0.5% to 4.0% of all cases of Crohn’s disease [1, 2]. Isolated duodenal Crohn’s disease accounts for only 0.07% of all cases [2]. The diagnosis of CD depends on the clinical, laboratory, radiological, macroscopic and microscopic features. However, findings of granuloma are considered the hallmark of histological diagnosis, but it can be detected in only 15-36% of biopitic samples [11] and in only 40-60% in surgically resected bowel segments in patient's patients with CD [12].

2. Method

2.1. Background

The first report on duodenal CD was done by Gottlieb and Alpert in 1937. Since then, few case reports or small series have been reported. Therefore, the diagnosis may be delayed,
and optimal treatment practices remain poorly defined [3, 4].

We encountered a case of a 55-year-old woman and diagnosed her with isolated duodenal CD and associated H Pylori gastritis following presentation with upper abdominal pain, early satiety and weight loss. There was no clinical evidence of involvement of the other small bowel segments or colon. Here we describe the details of this case with a review of the literature.

We think this case is important because of the relatively high prevalence of Crohn’s disease in general but very rare to have it only in the duodenum. That needs more suspicion and further work up.

2.2. Case Presentation

A previously healthy 55 - year- old lady was referred to the gastroenterology team with a working diagnosis of possible malignant gastric outlet obstruction and thickened duodenal wall on abdominal imaging following admission with upper abdominal pain, early satiety and weight loss (7 kg in 4 months). There were no provoking or palliating factors. Her symptoms were not responding to Proton pump inhibitors (PPI). She had no diarrhea nor rectal bleeding. Systemic review was normal and specifically she reported no joint pain, eye problems or skin condition. She had gastroscopy and colonoscopy four years prior to her presentation to investigate iron deficiency anaemia and reported to have normal looking oesophagus, stomach and mild duodenitis. Duodenal biopsy reported normal duodenal mucosa and no evidence of coeliac disease. Ileocolonoscopy and biopsies were normal. Her son was known to have CD and her uncle had esophageal cancer. She did not drink alcohol or smoke and no history of non-steroidal anti-inflammatory drugs (NSAID) or Aspirin use. She was post-menopausal by 3 years.

The physical examination was unremarkable. Her laboratory tests revealed iron deficiency anaemia (Table 1). Inflammatory markers were normal as were renal and liver functions. She had Computed Tomography (CT) of thorax, abdomen and pelvis which showed significant duodenal wall thickening up to 7.4mm for approximate length of 36mm (about 1.42 in) at the level of D1/D2 junction with narrowing of the lumen up to 3.5mm (Figure 4). No evidence of complete obstruction or any sinister lesion. The rest of the small bowel and the colon were normal. Subsequently, she underwent repeat OGD and biopsies. The OGD showed infiltrated edematous mucosa of the duodenal bulb with severe stenosis at the junction between the first part of the duodenum (D1) and the second part (D2), lumen less than 6mm (Figure 2). Multiple duodenal biopsies were obtained. They showed heavily acutely inflamed duodenal mucosa with focal ulceration. There was no evidence of dysplasia or malignancy. A diagnosis of severe acute ulcerative duodenitis with partial gastric outlet obstruction was made, and subsequently endoscopic dilatation was performed (Figure 3). Her colonoscopy was performed up to the terminal ileum and showed no significant findings. The biopsies from throughout the colon and terminal ileum were normal. She was kept on double dose PPI but despite that her symptoms persisted.

3. Result

Given the high index of suspicion of possible duodenal Crohn’s disease, the patient had repeat gastroduodenoscopy three months later which revealed the same findings of mucosal oedema with serpentine-like ulcerations at the duodenal bulb and partial stenosis at D1/D2 junction (Figure 2). More dilatation was done, and extensive biopsies were taken. This time the duodenal biopsy detected non-caseating epithelioid granulomas, features were in keeping with Crohn’s disease of the duodenum (Figure 5). The gastric biopsies showed Helicobacter Pylori (H. Pylori) gastritis. According to the above significant findings, the diagnosis of isolated duodenal CD was confirmed in addition to Pylori gastritis. Eradication therapy with pantoprazole, Clarithromycin and Amoxicillin was given for two weeks. The patient started on a short course of tapered prednisolone 40mg and commenced biological treatment with infliximab infusion following negative prebiological workup.

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
<th>Reference value</th>
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<tbody>
<tr>
<td>WBC (/L)</td>
<td>4.7</td>
<td>4 × 10⁹ - 10 × 10⁹</td>
</tr>
<tr>
<td>Hb (g/dl)</td>
<td>11.7</td>
<td>12 - 15</td>
</tr>
<tr>
<td>MCV (fl)</td>
<td>73.8</td>
<td>83 - 101</td>
</tr>
<tr>
<td>MCH (pg)</td>
<td>22.9</td>
<td>27 - 32</td>
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<tr>
<td>MCHC (g/dl)</td>
<td>31.0</td>
<td>31.5 - 36.0</td>
</tr>
<tr>
<td>Platelets (/L)</td>
<td>325</td>
<td>150 × 10⁹ - 400 × 10⁹</td>
</tr>
<tr>
<td>Iron (umol/L)</td>
<td>5.2</td>
<td>6.6 - 26</td>
</tr>
<tr>
<td>Transferrin (g/L)</td>
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<td>2.0 - 3.6</td>
</tr>
<tr>
<td>TF Saturation (Calc)</td>
<td>6.7</td>
<td>20 - 50</td>
</tr>
<tr>
<td>Ferritin (ug/L)</td>
<td>8</td>
<td>13 - 150</td>
</tr>
<tr>
<td>CRP (mg/L)</td>
<td>0.7</td>
<td>0 - 5</td>
</tr>
<tr>
<td>Blood urea (mmol/L)</td>
<td>4.2</td>
<td>2.5 - 7.8</td>
</tr>
<tr>
<td>Creatinine (umol/L)</td>
<td>83</td>
<td>44 - 80</td>
</tr>
</tbody>
</table>

WBC: White blood cell count; Hb: Haemoglobin; MCV: Mean corpuscular volume; MCH: Mean corpuscular haemoglobin; MCHC: Mean corpuscular haemoglobin concentration; CRP: C-reactive protein.

Figure 1. Normal looking colonoscopy at the terminal ileum.

Table 1. Laboratory Data.
The patient had marked clinical response with resolution of all symptoms. Repeated gastroscopy at 6 months on Infliximab infusions showed complete endoscopic remission with residual minimal fibrotic stenosis at D1/D2 junction (Figure 6). Histopathology confirmed mucosal remission and gastric biopsies revealed no H. Pylori.

4. Discussion

Crohn’s disease is a chronic and inflammatory disease characterized by the segmented, transmural involvement of the alimentary tract that can affect any part of the system from the mouth to the anus [5]. Patients with Duodenal Crohn’s disease (DCD) usually present with Crohn’s disease affecting other areas of the gastrointestinal tract; however, isolated DCD is a rare clinical entity of which diagnosis requires a high level of clinical suspicion [6]. This patient presented with duodenal stricture which can occur from a range of benign infective - inflammatory to malignant etiology. Many pathologies in the duodenum (both intrinsic and extrinsic) can cause duodenal obstruction [13]. Their etiologies are diverse and depend on the patient’s age [14]. The main causes in infancy are duodenal atresia, duodenal webs, and annular pancreas. In adult patients’ peptic ulceration, inflammatory strictures, trauma and malignancy (primary or secondary) are more common.

Initially, patients with DCD are managed with a combination of antiacid and immunosuppressive therapy. However, medical treatment fails in the majority of DCD patients, and surgical intervention is required in case of complicated disease. The most common indication for surgical intervention is progressive obstruction, failure of medical management with intractable pain, bleeding, perforation, and fistulous disease [7, 8].

Although Crohn’s disease of the upper gastrointestinal tract is negatively associated with H. Pylori, the detection of non-caseating granuloma helped diagnose Crohn’s disease. A study from Korea showed that 25% was associated with Pylori positive gastritis in comparison to 40% association with Pylori negative gastritis [15]. The patient coeliac markers were negative, and the duodenal biopsies did not show any features of coeliac.

The treatment of this patient structuring disease involved a multifactorial approach including possible early surgical intervention, total parenteral nutrition, steroids, dilations, and anti-tumor necrosis factor (TNF).

The treatment with Infliximab for isolated duodenal Crohn’s disease is limited to a few case reports with unknown long-term outcome [5, 9]. In a previous study 11 patients with Crohn’s disease that involved the upper gastrointestinal tract were treated with anti-TNF therapy (10 with infliximab and 1 with adalimumab). After 12 weeks, 8 of the 11 patients had significant macroscopic improvement and 7 had significant histologic improvement of their upper GI disease [10].

In this case the duodenal stricture did not respond to
dilation alone and repeated sampling was important to get the final diagnosis.

In summary, it is rare to have isolated duodenal CD. This case emphasizes that a high index of suspicion is necessary to diagnose isolated duodenal CD. It also highlights the many therapeutic challenges that we might face in severe stenosis or obstruction and a multidisciplinary approach would be beneficial.

5. Conclusion

Symptomatic duodenal CD is an uncommon disease presentation, especially in isolation. The most common duodenal disease phenotype is stricturing disease rather than inflammatory or perforating.

The majority of patients with duodenal CD have concurrent involvement of the terminal ileum or large intestine at presentation, rather than presenting with symptomatic CD of the duodenum alone as in our case.

In high index of suspicion of isolated duodenal Crohn’s disease, further endoscopic follow up and early repeat sampling are recommended for diagnosis.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Conflict of Interests

All authors declare that there is no conflict of interests to disclose in the study.

References


