A Rare Presentation of Esophageal Lichen Planus in a Patient with Collagenous Gastritis and Duodenitis: A Case Study

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Introduction

Esophageal Lichen Planus (ELP) is an atypical and potentially underdiagnosed cause of esophagitis and refractory strictures, most commonly presenting in middle to elderly aged women. We report a rare case highlighting isolated ELP in a patient with concomitant collagenous gastritis (CG) and duodenitis (CD).

Case Presentation

- An 80-year-old Caucasian woman was evaluated for refractory esophageal stenosis associated with dysphagia, loss of appetite and a 30-pound weight loss.
- Patient denied altered bowel habits or associated skin lesions.
- Past medical history: arthritis, hypertension, hypercholesterolemia, cystitis, fibromyalgia, skin cancer and history of chicken pox, measles and rheumatic fever
- She had undergone multiple endoscopic dilations as well as a gastrotomy tube placement due to poor nutrition, with minimal improvement.
- Biopsies from prior endoscopies were suggestive of erosive/ulcerative esophagitis.

Intervention

- Patient underwent endoscopy (Figure 1) which revealed a narrow caliber esophagus lined by sloughing mucosa and a normal gastroesophageal junction. The stomach and small bowel mucosa appeared atrophic and pale.
- Biopsies of the esophagus depicted Civatte bodies and characteristic lymphocytes consistent with lichen planus esophagitis.
- Duodenum and stomach bowel biopsies (Figure 2, Figure 3) depicted denudation of villi and surface epithelium, hyalinized subepithelial collagen deposition and chronic inflammatory infiltrate in the lamina propria, consistent with CG and CD.
- Patient was started on once-daily 3mg swallowed budesonide slurry which markedly improved her dysphagia and helped regain weight.

Endoscopic Findings

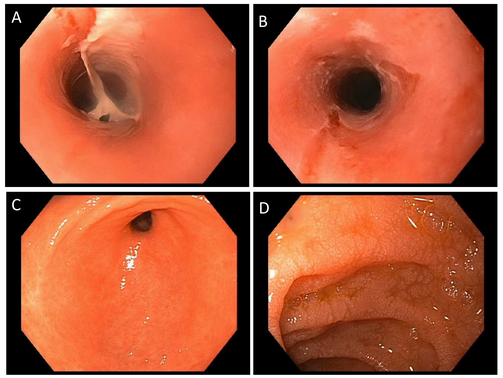

Figure 1. (A) Sloughing and edematous esophageal mucosa; (B) Edematous narrow caliper esophagus with erosions; (C) Atrophic appearing mucosa in gastric antrum; (D) Flattened villi in second portion of duodenum.

Histopathological Findings

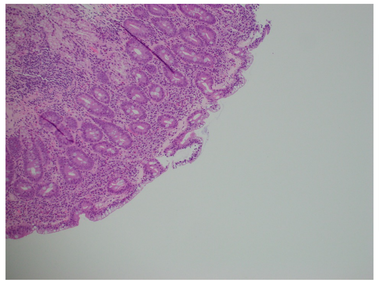

Figure 2. Histological findings of collagenous duodenitis. Duodenum depicting signs of villous atrophy, subepithelial collagen deposition with background chronic inflammation.

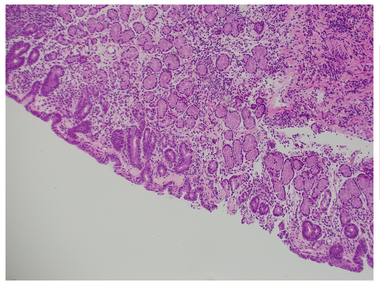

Figure 3. Histological findings of collagenous gastritis. Gastric antrum depicting subepithelial collagen deposition with background chronic inflammation.

Discussion

- The prevalence of ELP is estimated to be 0.19%; however, may be underdiagnosed since routine endoscopy is not performed in all LP patients.[1]
- Endoscopic findings can include sloughing edematous mucosa, web-like strictures, white exudate and pseudomembranes often affecting the proximal esophagus with typical sparing of the gastroesophageal junction.[1-4]
- The etiology has been postulated to involve a cell-mediated immune response where $CD8^+$ cytotoxic T-cells target basal keratinocytes producing an inflammatory response destroying the surrounding mucosal epithelium.[1,3]
- ELP patients have been noted to have an increased incidence of thyroid dysfunction.[5]
- Malignant transformation of ELP to esophageal squamous cell carcinoma has been reported.
- No standardized treatment regimen exists for ELP, but clinical improvement has been demonstrated with the use of systemic or oral corticosteroids, retinoids, or immunosuppressants like cyclosporine or azathioprine.[1-3]

- The collagenous gastroenteritides are an uncommon group of inflammatory disorders characterized by similar histopathologic findings with simultaneous occurrence in most patients.
- CG and CD are rare entities often associated with concurrent collagenous colitis, rather than being an isolated finding.
- CG demonstrates a female predominance and bimodal age distribution, typically presenting with symptoms of abdominal pain, anemia, diarrhea and weight loss.[6-8]
- Pathogenesis has been postulated to involve type III and IV collagen deposition as a restorative response secondary to previous inflammatory, infectious or toxic injury.[6]
- Adult collagenous gastroenteritides have been associated with autoimmune disorders – including celiac disease, collagenous sprue, lymphocytic gastritis and colitis, and inflammatory bowel disease.[6-9]
- No standard treatment protocol exists, but clinical improvement has been demonstrated with corticosteroids, proton pump inhibitors, H2-receptor antagonists, iron supplementation, diet modification, among others.

Conclusions

- There should be a high index of suspicion for ELP in patients with the correct demographics presenting with dysphagia, proton pump inhibitor (PPI) refractory esophagitis and stenosis.
- Endoscopic evaluation should be considered in LP patients to define disease extent – as many cases are believed to go undiagnosed due to absent or variable symptomatology that can mimic other conditions.
- Additional evaluation and biopsies should be considered if refractory symptoms or those consistent with another disease process are present.
- The lack of literature and scant incidence of both conditions suggests that further analysis of ELP patients should be considered to determine if there is an association with CG or CD, as this may also shed light on their associated pathogenesis.

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