Small Bowel Imaging in Celiac Disease

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KEYWORDS

• Enteroscopy • Small intestine • Enterography • Imaging • Celiac disease

KEY POINTS

- Recent advances in small bowel imaging technologies have improved the care of patients with small bowel diseases. Small bowel endoscopic and radiologic technologies are complementary and are often used in conjunction.
- In patients with celiac disease and gastrointestinal symptoms, radiologic imaging may be diagnostic of celiac disease.
- It is critical that radiologists and gastroenterologists are familiar with findings suggestive of celiac disease with new imaging modalities.
- Video capsule endoscopy, enterography, and device-assisted enteroscopy are usually reserved for those with alarm symptoms, refractory celiac disease, or suspicion of small bowel lymphoma/adenocarcioma.

INTRODUCTION

The small intestine is the longest organ of the gastrointestinal tract, which is fanned on the mesenteric stalk in the abdominal cavity. The multiple folds of small bowel loops in the abdominal cavity and peristalsis make it difficult to examine using standard endoscopic and radiologic imaging techniques. As a result, the small intestine has long been considered the black hole of the gastrointestinal tract. Upper endoscopy, push enteroscopy, and colonoscopy allow examination of only a small portion of the jejunum and distal ileum. Radiologic imaging by small bowel barium study and traditional abdominal computed tomography (CT) show luminal findings suggestive of celiac disease but provide poor examination of the small bowel wall. In addition, small bowel series is a time-consuming, operator-dependent study that has fallen out of favor for CT imaging. Recent advances in endoscopic imaging technologies (capsule and device-assisted eneroscopy) have enabled detailed visualization of the

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entire small bowel mucosa. Radiologic advances (CT enterography [CTE] and magnetic resonance enterography [MRE]) have markedly improved examination of the small bowel wall and surrounding structures. Intraoperative enteroscopy, considered the gold standard of complete enteroscopy, has largely been replaced. New imaging technologies provide less-invasive methods, with excellent reader agreement, for examining the small bowel. These technologies have improved the diagnosis and management of patients with small bowel diseases, such as small bowel bleeding, Crohn, disease, and celiac disease.

Celiac disease is a common inflammatory disease of the small bowel that affects 1% of the white population.[1] It is triggered, in genetically predisposed individuals, by the ingestion of gluten, a protein component of wheat, rye, and barley.[2] Most individuals are diagnosed with celiac disease in adult life. Celiac disease may present with classic (diarrhea-predominant) symptoms or atypical symptoms or may be asymptomatic and detected via screening.[3] Atypical presentations of celiac disease include nonspecific abdominal pain/dyspepsia, constipation, bloating, reflux, infertility, anemia, osteoporosis, dental enamel defects, short stature, vitamin deficiencies, fatigue, or neurologic problems, such as neuropathy or ataxia.[4] The rash of dermatitis herpetiformis is virtually always associated with celiac disease whereas intestinal biopsy may be normal with gluten ataxia. Serology for the antibodies directed against tissue transglutaminase (tTG IgA) is the best screening test for celiac disease.[5] The gold standard for diagnosis is upper endoscopy with small bowel biopsy. Patients with positive serology should have an upper endoscopy performed with 4 to 6 biopsies of the small intestine with samples from both the bulb and the second portion of the duodenum to maximize yield.[6,7] If the suspicion for celiac disease is high, endoscopy with biopsy should be performed despite negative serology results. Endoscopic findings of celiac disease include loss of folds and scalloping, a mosaic pattern, and fissuring of mucosa (**Fig. 1**).[8,9] Endoscopic abnormalities are not present in all cases and biopsies should be obtained even if the duodenum appears normal. Marsh[10] and Oberhuber and colleagues[11] described the histologic changes of celiac disease as increased intraepithelial lymphocytes, crypt hyperplasia, and villous atrophy. These findings largely account for abnormalities detected on radiologic imaging studies. Endoscopic biopsies of the duodenum may be normal if the disease is patchy. Jejunal biopsies may improve diagnostic yield in such patients.

The only current available treatment of celiac disease is a gluten-free diet. Up to 30% of celiac patients experience continued symptoms on a gluten-free diet or incomplete histologic recovery and are considered nonresponsive.[12,13] Other causes of ongoing symptoms include continued gluten exposure, microscopic colitis, small intestinal bacterial overgrowth, lactose or fructose intolerance, pancreatic exocrine insufficiency, and refractory celiac disease (RCD).[14] RCD is defined as persistent diarrhea with villous atrophy, crypt hyperplasia, and inflammation, despite adherence to a strict gluten-free diet for 6 to 12 months.[15,16] Its prevalence is unknown. RCD is classified into type 1 and type 2 depending on intraepithelial lymphocyte expression.[17] In RCD type 1, intraepithelial lymphocytes have normal surface expression of both CD3 and CD8 with a polyclonal T-cell receptor. In RCD type 2, however, an abnormal lymphocyte population is present with a loss of surface CD3 and CD8 expression, retention of intracellular CD3, and a monoclonal T-cell receptor rearrangement. In a study of 57 patients with RCD, the 5-year survival rate was 93% in patients with RCD type 1 and 44% with RCD type 2.[18] Enteropathy-associated T-cell lymphoma (EATL) occurs in more than 50% of patients with RCD type 2 and is a significant cause of mortality.[17–20] Evaluation of poorly responsive and refractory celiac patients includes consultation with a skilled dietician, endoscopy with biopsy, stool studies,