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캡슐 내시경으로 평가된 만성 비육아종 궤양성 공회장염

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Chronic Non-granulomatous Ulcerative Jejunoileitis Assessed by Wireless Capsule Endoscopy

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Chronic non-granulomatous jejunoileitis is a rare disease characterized by malabsorption, abdominal pain, and diarrhea that causes shallow ulcers in the small bowel. The etiology of chronic non-granulomatous jejunolieitis remains unknown. A 69-year-old man complained of abdominal pain and lower extremity edema. A 99m-Tc albumin scan showed increased radioactivity at the left upper quadrant, suggesting protein-losing enteropathy. A small bowel follow-through did not disclose any lesions. Wireless capsule endoscopy revealed several small bowel ulcers and strictures. A jejunoileal segmentectomy with end-to-end anastomosis was performed, and the histologic examination revealed non-granulomatous ulcers with focal villous atrophy. Ruling out all other possible diagnoses, we diagnosed our patient with chronic non-granulomatous ulcerative jejunoileitis. Postoperatively, the patient's abdominal pain and lower extremity edema improved, and the serum albumin normalized. This is the first case of chronic non-granulomatous ulcerative jejunoileitis localized by wireless capsule endoscopy and treated successfully with segment resection. **(Korean J Gastroenterol 2010;56:382-386)**

Key Words: Ileitis, Ulcerative; Hypoalbuminemia; Malabsorption syndromes; Capsule endoscopy

Introduction

Chronic non-granulomatous ulcerative jejunoileitis is a rare disease of unknown etiology. This rare disease presents with chronic diarrhea, weight loss, and malabsorption.[1] The inflammatory process may range from mucosal inflammation to the involvement of the deeper bowel layers, resulting in the ulceration and narrowing of the jejunum and ileum.[2] Because chronic non-granulomatous jejunoileitis involves the small bowel, it is difficult to examine the lesions using conventional techniques. The development of wireless capsule endoscopy (WCE) has facilitated the exploration of the small bowel by providing direct images. WCE can target the suspected small bowel lesions and minimize the use of invasive procedures.[3]

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This is the first report demonstrating that WCE plays an essential role for detecting chronic non-granulomatous ulcerative jejunoileitis and localizing the pathologic segment.

Case Report

A 69-year-old man had lower extremity edema for 3 years and a 10-year history of intermittent postprandial abdominal pain. Hypoalbuminemia was detected 3 years ago and treated intermittently with intravenous albumin. However, pitting edema of the lower extremities and abdominal pain worsened as time went on. He had no medication history including NSAIDs (nonsteroid anti-inflammatory drugs). The laboratory findings showed remarkable hypoproteinemia and hypoalbuminemia with normal liver and renal function; the serum protein was 3.9 g/dL and the serum albumin was 1.6 g/dL. The hemoglobin was 12 g/dL, the white blood cell count was 6,300/mm3, and the platelet count 378,000/mm^3. The stool was positive for fecal occult blood at 1,149 ng/mL. The serum IgA, IgM, and IgG levels were normal. Protein-losing enteropathy was highly suspected; however, α1-antitrypsin clearance could not be checked due to test unavailability. Instead, a 99mTc-albumin scan showed increased radioactivity at the left upper quadrant (Fig. 1). Esophagogastroduodenoscopy and colonoscopy were reported to be normal, and multiple biopsies from stomach, duodenum, terminal ileum, and colon showed no pathologic findings. An abdominopelvic CT scan revealed no abnormal findings. To evaluate the suspected small bowel lesions, a small bowel follow-through was performed. However, the results were unremarkable. To examine the suspected small bowel lesions more precisely, WCE (PillCam^{TM} SB; Given Imaging, Yoqneam, Israel) was performed, and multiple mucosal erosions and shallow ulcers with villous atrophy were visualized in jejunoileal lesion (Fig. 2). In addition, stenotic foci led to WCE retention without obstructive symptoms. For proper diagnosis and treatment, 34.5 cm of the jejunum and ileum was resected with an end-to-end anastomosis. Intraoperative enteroscopy was done to confirm the disease extent. The gross specimen showed a dilated proximal bowel segment 4.2 cm in diameter. The mesenteric fat tissue was mildly adherent to the serosal surface focally with no fibrous adhesion bands, creeping fat, tumefaction, or enlarged tumorous lymph nodes (Fig. 3A). On opening, several shallow transverse or oval erosions to ulcerations, ranging from 1.2×1.1 to 5.4×1.1 cm, were observed (Fig. 3B). Two white fibrous stenotic foci were noted without

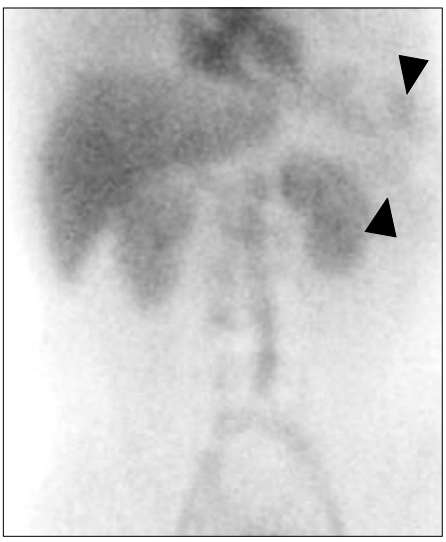

Fig. 1. 99m-Tc albumin scan finding. At 2 hours, increased radioactivities at the left upper quadrant (arrowheads) were noticed.

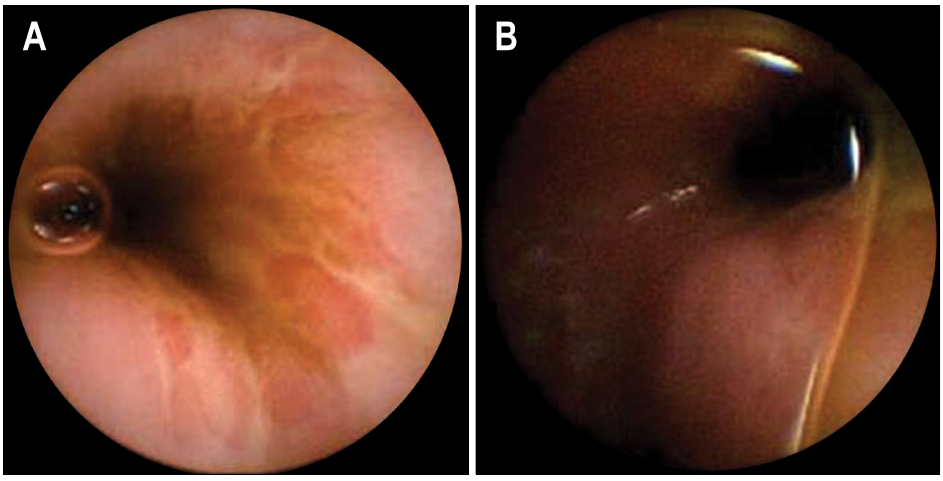

Fig. 2. Wireless capsule endoscopy findings. (A) It revealed multiple geographic and shallow ulcerations with villous atrophy on the distal jejunum. (B) It showed stenotic area, which caused capsule retention in the proximal ileum.

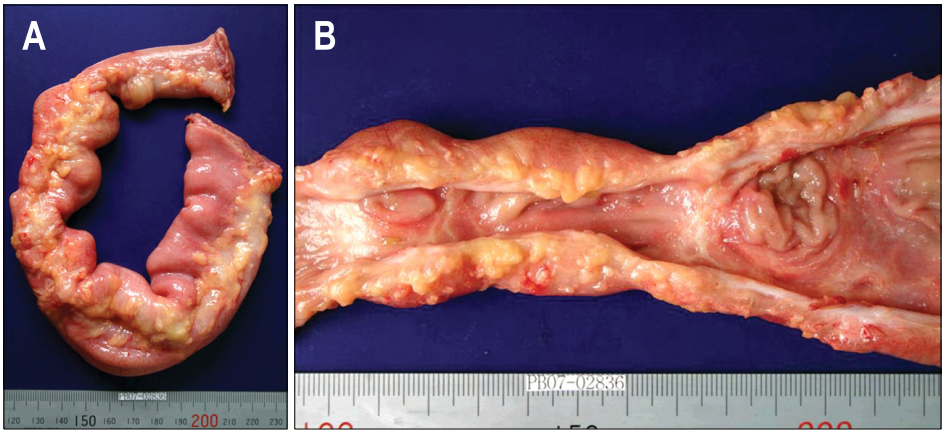

Fig. 3. The gross findings of resected specimen. (A) It showed a dilated proximal bowel segment 4.2 cm in diameter. The mesenteric fat tissue was mildly adherent to the serosal surface focally. (B) On opening, several shallow transverse or oval ulcerations were observed with two white fibrous stenotic foci.

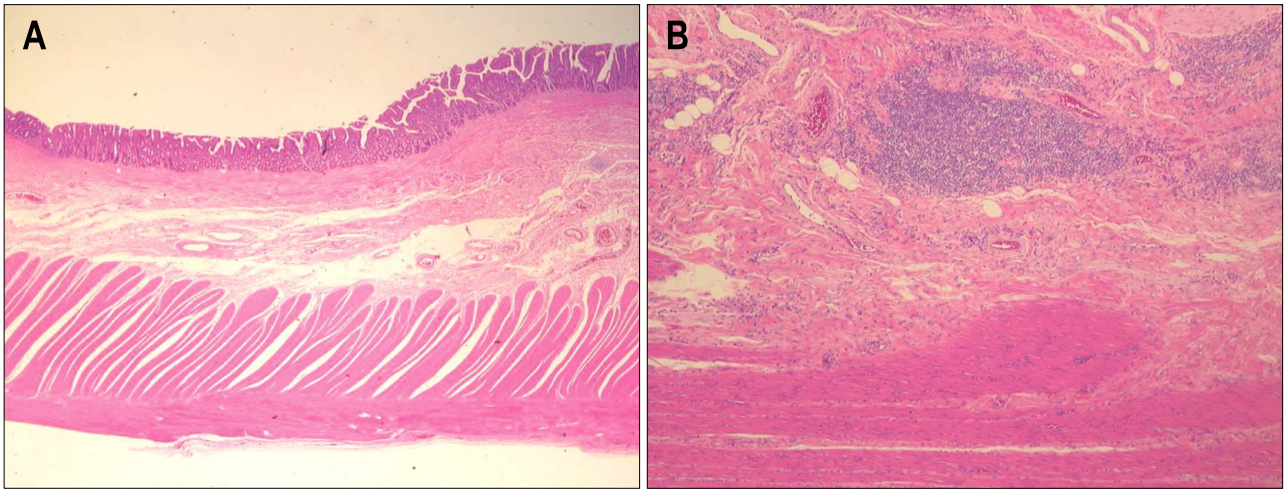

Fig. 4. Microscopic findings. (A) It showed non-granulomatous ulcers with focal villous atrophy (H&E, ×40). (B) A few cryptitis foci were observed (H&E, ×100).

obstruction (Fig. 3B) and the WCE was found in the proximal stenotic area. Microscopically, non-granulomatous ulcers with focal villous atrophy were observed (Fig. 4A). A few cryptitis foci were observed, but no crypt abscesses were present (Fig. 4B). No lymphocytic vasculitis, microorganisms, parasitic ova or worms, cytomegalopathy, or viral inclusions were observed. Surgical resection of the jejunoileal ulcers and strictures resulted in clinical remission. Post-operatively, the serum albumin increased steadily to 3.6 g/dL, and the patient's abdominal pain resolved. He has had no further symptoms related to chronic non-granulomatous ulcerative jejunoileitis.

Discussion

Given the histologic and clinical similarities, chronic non-granulomatous jejunoileitis is often considered to be a form of gluten-sensitive enteropathy. In many cases, the disorder develops in compliant celiac patients with stable disease, whereas in other cases, the disorder presents *de novo* in people with no known pre-existing disease.[4] Since Nyman's original report in 1949, 70 cases of patients with intestinal inflammation and ulcerations have been reported. Thirty six patients (51%) had underlying disease; of these, 23 (64%) had celiac sprue, 10 (29%) had intestinal lymphoma, and 5 (14%) had hypogammaglobulinemia. Thirty four patients did not have underlying disease causing chronic nongranulomatous jejunoileitis.[4] On histopathologic findings, our case showed non-granulomatous ulcers with focal villous atrophy. A few cryptitis foci without crypt abscesses were also observed. Although focal villous atrophy was present around the ulcers, the typical change in the villous epithelium was absent,[5] and the duodenal biopsies did not reveal any villous epithelial change or villous atrophy.

Therefore, we excluded the diagnosis of celiac sprue. In addition, celiac sprue has never been reported in Korea. We also considered cryptogenic multifocal ulcerous stenosing enteritis (CMUSE) in the differential diagnosis. CMUSE is characterized by intermittent bouts of intestinal obstruction, ulcerative stenoses of the small bowel relapsing after surgical resection, and steroid sensitivity. Unlike chronic non-granulomatous ulcerative jejunoileitis, CMUSE has a normal villous architecture and no malabsorption.[6] In our case, the microscopic examination showed focal villous atrophy, and obvious hypoalbuminemia was present, ruling out CMUSE. We concluded that our patient had idiopathic non-granulomatous ulcerative jejunoileitis after ruling out other possible pathologic conditions, such as Crohn's disease, tuberculosis, fungal infections, corticosteroid therapy, intestinal lymphoma, and celiac sprue.[4]

Chronic non-granulomatous jejunoileitis is a rare disease entity with unknown etiology, typically presenting with abdominal pain, diarrhea, and malabsorption. The usual findings of chronic non-granulomatous ulcerative jejunoileitis are small bowel thickening or edema and hyperemia of the serosa, most often of the jejunum. Multiple strictures with proximal intestinal dilatation are common. Ulcers are typically superficial and often perpendicular to the long axis of the bowel,[7] as in our case. Most cases remain undiagnosed until laparotomy, which is often precipitated by complications, such as obstruction, hemorrhage, or perforation.[7] Our patient had chronic abdominal pain and lower extremity pitting edema due to hypoalbuminemia, despite intermittent albumin infusions. The 99m-Tc-albumin scan showed increased radioactivity at the left upper quadrant, suggesting protein-losing enteropathy, and the fecal occult blood test was strongly positive. However, we were unable to verify lesions on small bowel follow-through. WCE is known to facilitate the detection and assessment of ulcerated mucosal lesions located in the small bowel.[8] We confirmed the ulcerative and stenotic lesions by WCE. Three patients with chronic non-granulomatous jejunoileitis have been reported in Korea, and their manifestations were similar to the manifestations in our patient: (abdominal pain, protein-losing enteropathy, and anemia).[1] Nevertheless, it is still very rare, and there are no reports of chronic non-granulomatous jejunoileitis detected and localized by WCE.

While examining the lesions, the WCE was retained in a stenotic segment. Capsule retention is the most common complication of WCE.[9] Prophylactic measures to minimize capsule retention have been disappointing.[10] Moy and Levine reported that a retained capsule in the small intestine can occur with a normal barium study before performing WCE,[11] and the current version of the patency capsule system (Agile Patency System; Yoqneam, Israel) fails to add substantial additional information about the risk of capsule retention compared with radiographic imaging modalities.[12] Capsule retention usually does not cause obstructive symptoms,[13] and in our case, he neither experienced obstructive symptoms after capsule retention.

The prognosis of chronic non-granulomatous ulcerative jejunoileitis is usually poor; however, the localized form has a good prognosis after resecting the pathologic segments.[7,14] Resection of the ulcerated or stenosed bowel gives the best chance of prolonged survival. Our patient was categorized as the local form and underwent resection of the pathologic lesion, which caused capsule retention and recovered from this illness successfully.

In conclusion, this is the first report in which chronic non-granulomatous ulcerative jejunoileitis was detected and localized by using WCE. We experienced the accurate localizing ability of WCE for ulcerative small bowel lesions, which caused protein loss and was identified as chronic non-granulomatous jejunoileitis.

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