Case 1: Protein-losing Enteropathy with Generalized Warts

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Introduction

Primary intestinal lymphangiectasia (PIL) is a congenital and rare disorder characterized by dilated intestinal lymphatics resulting in lymph leakage and protein-losing enteropathy.\(^1\) Due to loss of lymphocytes, especially CD4\(^+\) T cells, PIL is associated with cell mediated immunodeficiency.\(^1\) As a result, the patients are vulnerable to chronic viral infection and lymphomas. However, PIL with chronic viral infection such as human papilloma virus-induced generalized warts was very rarely reported.\(^1\)\(^-\)\(^5\) Capsule endoscopy can be useful tool for diagnosing PIL through examination of the entire small intestine.

Fig. 1. He presented with generalized warts over his entire body, including both hands and feet.
Case Report

A 36-year-old man was admitted to the hospital with a 3-month history of diarrhea and weight loss (5 kg). He presented with generalized warts over his entire body, including both hands and feet (Fig. 1) with an initial onset at age 10. Family history was not remarkable. Laboratory tests showed hypoalbuminemia (albumin, 2.3 g/dL), hypogammaglobulinemia (IgG, 653.4 mg/dL), lymphopenia (843/mm³, CD4⁺ T cells, 24.4%; CD3⁺ T cells, 54.7 mg/dL) and increased stool a‐1 antitrypsin clearance (220.11 mL/24hr). Upper endoscopy showed diffuse mucosal edema in the duodenum. Colonoscopy revealed white mucosal plaques and spots in the terminal ileum and diffuse mucosal edema in the colon. Capsule endoscopy showed diffuse multifocal white mucosal plaques from the proximal jejunum to the terminal ileum, which was compatible with intestinal lymphangiectasia (Fig. 2). On histologic examination of the terminal biopsy specimens, H&E stain showed dilated lymphatic vessels with many foamy macrophages in lamina propria consistent with lymphangiectasia (A,×100), and CD34 stain showed normal vascular endothelial cells (B). Also found were D2-40 stained endothelial cells (red arrow), which indicated dilated lymphatics (C). Macrophages were observed by CD68 stain, which aggregated to uptake lipids leaking from dilated lymphatics (D).
minal biopsy specimens, H&E stain showed dilated lymphatic vessels with many foamy macrophages in lamina propria consistent with lymphangiectasia, and CD34 stain showed normal vascular endothelial cells. Also found were D2-40 stained endothelial cells, which indicated dilated lymphatics. Macrophages were observed by CD68-stain, which aggregated to uptake lipids leaking from dilated lymphatics (Fig. 3). Histological findings were also suggestive of PIL. Flow cytometry of peripheral blood lymphocytes showed reduced number of CD3^+ and CD4^+ T cells. He was ultimately diagnosed with PIL, and his warts were associated with T-cell mediated immunologic abnormalities. He was treated with a low fat/high protein diet with oral supplements.

**Discussion**

Diagnosis of PIL is not easy because abnormal lymphatic lesions usually distributed small intestine. Detection of lesions by upper gastroscopy and colonoscopy is limited, radiologic examinations such as computed tomography can’t confirm the diagnosis. In some cases, double balloon enteroscopy or surgical methods were used for pathologic examination. Biopsy can be performed by double balloon enteroscopy and it was also reported much higher diagnosis rate than capsule endoscopy. In our patient, the abnormal small intestine tissues were obtained in the terminal ileum by colonoscopic biopsy. Therefore, we performed capsule endoscopy in order to observe the entire small intestinal mucosa. Although capsule endoscopy can’t obtain biopsy, it is very useful method to examine mucosal change of small intestine. In our patient, capsule endoscopy showed multiple whitish spots, thickened villi and edematous mucosa of small intestine. These endoscopic findings are typical mucosal lesions of PIL. Therefore, capsule endoscopy is relatively easy, comfortable and safe method for diagnosing PIL.

**Conclusion**

We report a rare case of PIL with generalized warts diagnosed by capsule endoscopy.

**References**