

CASE REPORT

크론병과 연관되어 나타난 플러머-빈슨 증후군 1예

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A Case of Plummer-Vinson Syndrome Associated with Crohn's Disease

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Plummer-Vinson syndrome manifests as cervical dysphagia, iron deficiency anemia, an upper esophageal web, and atrophic glossitis. The cause of the esophageal web is thought to be iron deficiency anemia; however, the cause of Plummer-Vinson syndrome has not been established. Crohn's disease is usually accompanied by malnutrition and iron deficiency anemia; however, no case of concomitant Crohn's disease and Plummer-Vinson syndrome with aggravated malnutrition and anemia has been previously reported. Here, we report on a rare case of Plummer-Vinson syndrome in a Crohn's disease patient, which caused malnutrition and constipation. (*Korean J Gastroenterol* 2014;63:244-247)

Key Words: Plummer-Vinson syndrome; Malnutrition; Deglutition disorders; Crohn disease

INTRODUCTION

Plummer-Vinson syndrome is a clinical symptom complex of cervical dysphagia, iron deficiency anemia, upper esophageal web (or webs), and atrophic glossitis. Plummer documented 21 cases in 1912 and Paterson and Kelly defined its manifestation triad as: 1) cervical dysphagia, 2) iron deficiency anemia, 3) and esophageal web.¹ During the first half of the 20th century, Plummer-Vinson syndrome appeared to be common in the West; however, nowadays, the prevalence is low worldwide.² The syndrome is known to affect middle-aged females, and those affected may have other features of chronic iron deficiency, such as papillary atrophy of the tongue, spoon shaped brittle nails, angular stomatitis, and pica. In a series of 1,000 consecutive patients who underwent esophagography of the hypopharynx and cervical

esophagus, a web was found in 5.5% of cases. However, only six patients had dysphagia attributable to the presence of a web, and none of the patients fulfilled the criteria for Plummer-Vinson syndrome.^{1,2} The cause of esophageal web is uncertain; however, it is thought to be associated with iron deficiency anemia.

Crohn's disease (CD) is usually accompanied by malnutrition and iron deficiency anemia. However, no case of concomitant CD and Plummer-Vinson syndrome has been previously reported. Here, we report on a rare case of CD aggravation related to Plummer-Vinson syndrome.

CASE REPORT

A 34-year-old male visited our hospital due to severe general weakness with abdominal distension and pain. He had

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been diagnosed as having CD 15 years previously; however, he had not complied with prescribed medication for several years. He already had a history of repeated intestinal obstruction, and, because he refused surgery, had been placed on observation for several years.

He was admitted to the hospital due to dysphagia, constipation, severe malnutrition, and general weakness.

At presentation, he appeared to be chronically ill. Physical examination revealed abdominal distension due to ascites and pitting edema on both lower legs. No nail or tongue abnormality was noted. Laboratory findings were: white blood cell count $4,750/\text{mm}^3$, hemoglobin 8.9 g/dL (mean corpuscular volume 75 fL, mean cell hemoglobin 20 pg), hematocrit 30%, platelets $383,000/\text{mm}^3$, total protein 3.21 g/dL, albumin 1.0 g/dL, total bilirubin 0.43 mg/dL, AST 16 IU/L, ALT 7

IU/L, BUN 10.01 mg/dL, creatinine 0.97 mg/dL, Na 132 mEq/L, K 2.9 mEq/L, and PT 13.3 sec. Results of iron study for his anemia showed serum iron 45 μg , total iron-binding capacity 475 $\mu\text{g}/\text{dL}$, and ferritin 10 $\mu\text{g}/\text{L}$, which was suggestive of iron deficiency anemia. The patient's Crohn's disease activity index (CDAI) score on the day of admission was 230.3.

On the second hospital day, esophagogastroscopy (EGD) and sigmoidoscopy were performed for evaluation of his dysphagia symptoms. During EGD, a tight and narrow lumen that disturbed passage of the scope at the near upper esophageal sphincter (UES) was noted (Fig. 1). For further evaluation of the cause of passage disturbance, barium esophagography was performed and an upper esophageal web was observed (Fig. 2A, B). Sigmoidoscopy showed a cobble stone appearance with longitudinal ulceration at the sigmoid colon and a stenotic lumen was observed at 20 cm from the anal verge (Fig. 3). The scope could not be passed through this lesion. Abdominal computed tomography showed wall thickening at the terminal ileum and severe dilatation of the entire colon and deformity with luminal narrowing in the left side colon (Fig. 4).

According to laboratory findings, the patient had dysphagia, iron deficiency anemia, and endoscopy showed an upper esophageal web; therefore, we made a diagnosis of Plummer-Vinson syndrome.

We first attempted to remove the esophageal web with flexible upperendoscopy, but failed because the position of web was so close to UES, so that space for approaching flexible endoscopy was inadequate. Due to high risk of perforation through use of balloon dilatation or bougienage, we consulted to an otorhinolaryngologist for the dissection proce-

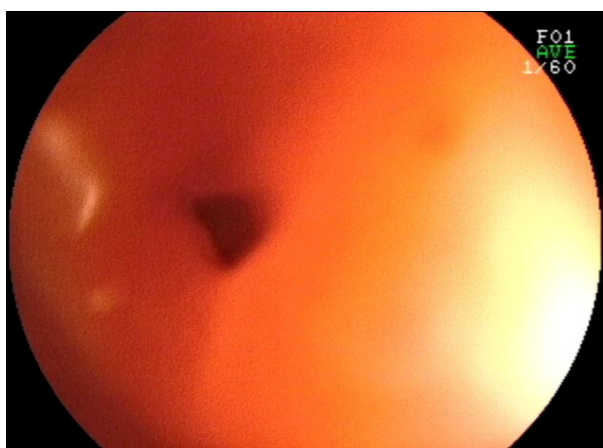


Fig. 1. Esophagogastroduodenoscopic finding. At the upper esophagus (near the upper esophageal sphincter) a white colored circumferential web was noted. The scope could not be passed through the lesion.

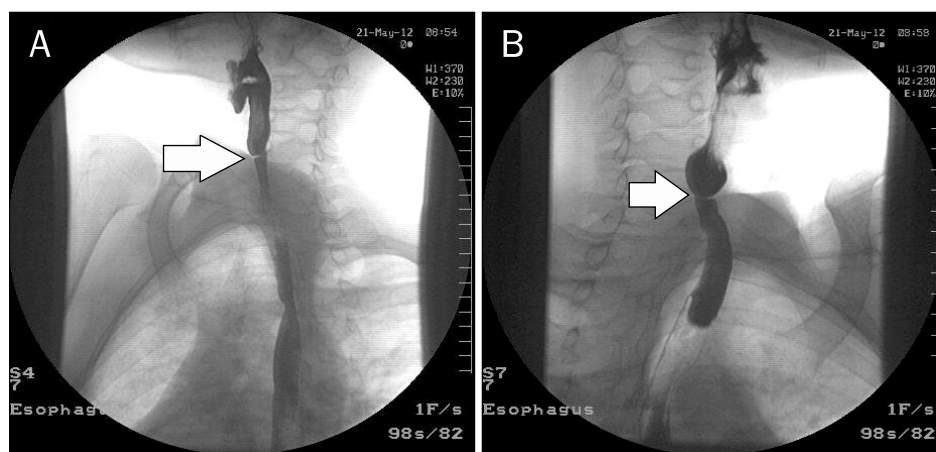


Fig. 2. Esophagography. A shelf-like filling suspicious for esophageal web defect was observed at the anterior wall with dilatation of the proximal part (A: frontal view, B: lateral view).

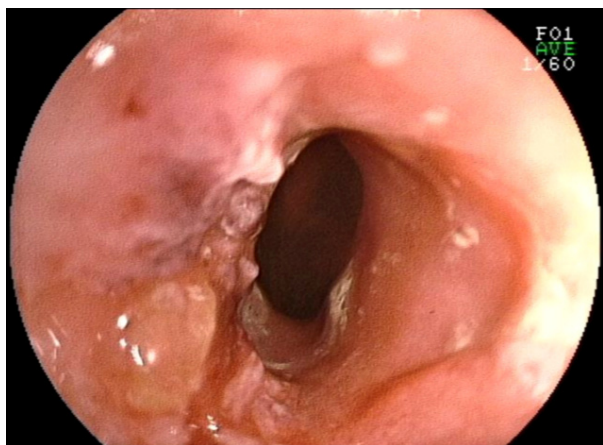


Fig. 3. Sigmoidoscopic finding. Cobble stone appearance with ulceration and luminal narrowing with stricture was noted at 20 cm from the anal verge.

dures using a rigid scope. On hospital day 5, for improvement of his dysphagia and nutritional problem, the upper esophageal sphincterotomy was dissected using a rigid scope and this procedure was performed under general anesthesia in the operation room and processed by an otorhinolaryngologist (admission day 5).

At the same time, a general surgeon performed resection of the stricture lesion of the sigmoid colon and made a colostomy site on the abdomen; this operation was performed immediately after esophageal web dissection.

The patient recovered his general condition without any specific complication after 10 days and he was able to eat a soft diet and also perform defecation easily. He was discharged 15 days after admission.

Three months after receiving this treatment, the patient was able to eat regular food and gained 10 kg in weight. The edema disappeared and serum albumin level was 3.2g/dL and his anemic state was eventually corrected to hemoglobin 12.0g/dL, hematocrit 42%, and CDAI score was 45.4. He has been in a remission state for one year after surgical correction of esophageal web and colon stricture.

DISCUSSION

The incidence of Plummer-Vinson syndrome is decreasing in the developed world, probably due to improved nutrition and the availability of health care systems.³

However, the precise incidence and prevalence of this syndrome cannot be easily determined. In addition, no case of



Fig. 4. Abdominal computed tomography. Ascites and terminal ileal wall thickening were observed. The arrow indicates one of the multiple strictures with prestenotic dilatation and bowel wall thickening at the left side colon.

concomitant CD with Plummer-Vinson syndrome has been previously reported.

The malnutrition and bowel lumen stricture caused by CD in our patient undoubtedly caused aggravation of iron deficiency anemia, which could have caused esophageal web formation or development of Plummer-Vinson syndrome, which, for diagnostic purposes, requires the presence of dysphagia, iron deficiency anemia, and one or more upper esophageal webs. These webs can be detected by esophagography or gastroendoscopy,¹ and are smooth, thin, and gray in appearance with eccentric luminal narrowing and a shelf-like structure. Typically, they occur in the upper part of the esophagus and can be missed or accidentally ruptured if an endoscope is not introduced under direct observation.¹

On the other hand, poorly controlled CD can result in colon stricture, which in turn, causes variable symptoms, including constipation, distended bowel, and dyspepsia, which aggravate malnutrition and iron deficiency anemia and also eventually provoke esophageal web formation and dysphagia.

Malnutrition, a common feature of CD, is caused by occult blood loss, mal-absorption at the intestinal surface due to inflammation, fistulae, edema, a surgically reduced intestinal surface area, or abdominal pain.⁴ In addition, nutritional status could affect the severity of inflammatory bowel disease. For example, some experts have suggested that optimal nutritional support can improve the status of patients with in-

inflammatory bowel disease and prevent the various complications of CD.⁵⁻⁸

The authors reported on a rare case of Plummer-Vinson syndrome associated with CD. The physician should also be aware of the importance of nutritional support in patients with inflammatory bowel disease.

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