

Relapsed Esophageal Web in a Patient with Plummer-Vinson Syndrome

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Plummer-Vinson syndrome is characterized by dysphagia, iron deficiency anemia, and upper esophageal web. The associated symptoms can be resolved by administering iron supplements as well as by endoscopic intervention. Relapse in patients with Plummer-Vinson syndrome is very rare. We describe a case of a 42-year-old woman with Plummer-Vinson syndrome whose symptoms were successfully treated with endoscopic dilatation and iron supplementation at first admission; however, 1 year later, she revisited our hospital because of dysphagia. On second admission, investigations revealed esophageal web relapse in Plummer-Vinson syndrome. She was again successfully treated with endoscopic dilatation and iron supplementation. After first admission, her anemia was not normalized due to poor compliance and loss of follow-up. We experienced a case of esophageal web relapse due to uncorrected iron deficiency anemia in a patient with Plummer-Vinson syndrome. This experience indicates that continuous iron supplementation and long-term follow-up is important in patients with Plummer-Vinson syndrome. (**Korean J Helicobacter Up Gastrointest Res 2014;14:117-120**)

Key Words: Plummer-Vinson syndrome; Iron deficiency anemia; Deglutition disorders; Recurrence

INTRODUCTION

Plummer-Vinson syndrome is rare, and involves the classic triad symptoms of dysphagia, iron-deficiency anemia and the presence of an esophageal web.^{1,2} Plummer-Vinson syndrome mostly occurs in women, with the highest incidence in the fifth decade of life.³ The pathogenesis of this syndrome remains unknown, with iron deficiency possibly being the most important etiologic factor.^{1,4} The prognosis of the syndrome is excellent, and patients can be treated easily with iron supplementation and mechanical dilatation.¹ Our search of the PubMed database indicated that cases of relapsed esophageal web in patients with Plummer-Vinson syndrome are very rare.

We report the case of a woman with Plummer-Vinson syndrome, in whom esophageal web relapse occurred because of uncorrected anemia; the patient's condition was successfully managed using endoscopic dilatation followed by iron supplementation.

CASE REPORT

A 42-year-old woman with a 3-year history of dysphagia presented to our hospital. She had anemia since delivery of her first child. On admission, laboratory data showed white blood cell count, 4,000/mm³ (normal range, 4,000~10,000/mm³); hemoglobin level, 6.0 g/dL (normal range, 12~16 g/dL); platelet count, 308,000/mm³ (normal range, 140,000~400,000/mm³); serum iron level, 13 µg/dL (normal range, 50~160 µg/dL); total iron binding capacity (TIBC), 363 µg/dL (normal range, 250~380 µg/dL); ferritin level, 1.44 ng/mL (normal range, 12~150 ng/mL); vitamin B₁₂ level, 841.3 pg/mL (normal range, 243~894 pg/mL); and folate level, 6.02 ng/mL (normal range, 3.1~17.5 ng/mL). Blood cell morphology showed microcytic hypochromic anemia, anisocytosis (++) and poikilocytosis (++) including elliptocytes, suggestive of iron deficiency anemia. Endoscopic examination demonstrated a post-cricoid esophageal web, through which the endoscope (Q260; Olympus, Tokyo, Japan) could not be passed (Fig. 1). A barium swallowing test revealed an upper esophageal web (Fig. 2). Based on these findings, a diagnosis of Plummer-Vinson syndrome was made. Endoscopic balloon

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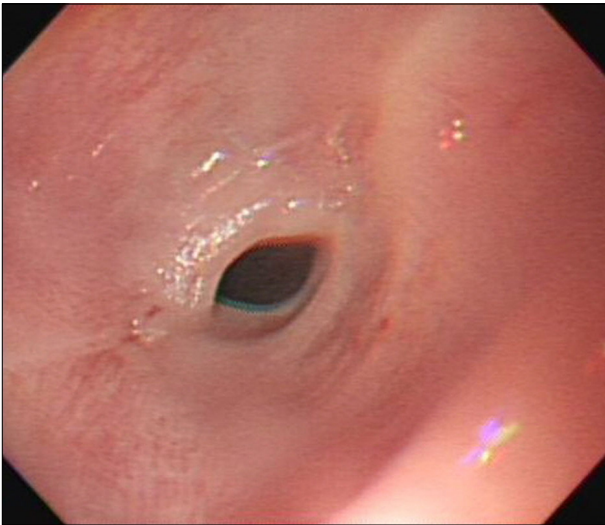


Fig. 1. Endoscopic findings at first diagnosis. An esophageal web observed at 17 cm from the incisor. The endoscope (Q260; Olympus, Tokyo, Japan) could not be passed through the web.

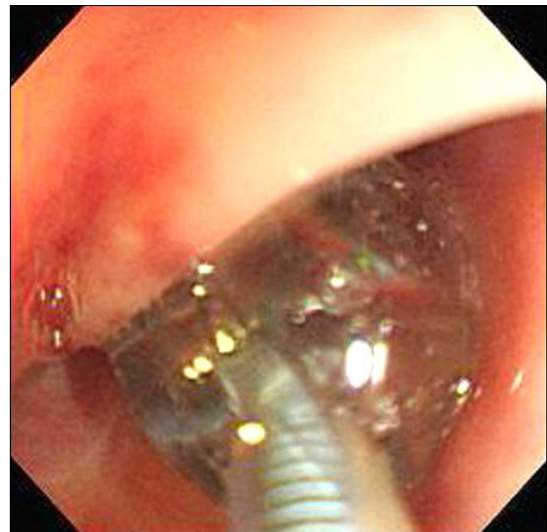


Fig. 3. Endoscopic findings during endoscopic treatment. Balloon dilatation at 8 mm and 10 mm for 1 min each.



Fig. 2. Barium swallowing test. Barium swallowing test revealed an upper esophageal web (arrow).

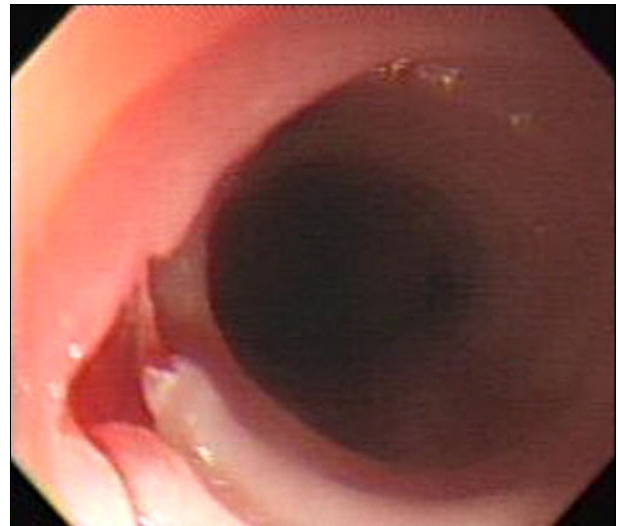


Fig. 4. Endoscopic findings after endoscopic treatment. After balloon dilatation, the endoscope (Q260; Olympus, Tokyo, Japan) could be passed through the web.

dilatation was performed (Fig. 3, 4), and iron supplementation was continued. Her symptoms resolved, and she was discharged. However, she was lost to follow-up. One year later, dysphagia recurred. On readmission, investigations revealed microcytic hypochromic anemia with hemoglobin level, 9.0 g/dL; serum iron level, 10 μ g/dL; TIBC, 339 μ g/dL; and ferritin level, 2.3 ng/mL. The endoscopic finding showed an upper esophageal web similar to that at the previous admission (Fig. 5). Therefore,

esophageal web relapse was diagnosed. Endoscopic balloon dilatation was performed again. After dilatation, her dysphagia resolved. At the time of discharge, the patient was explained the importance of taking iron supplement. After iron supplementation, no relapse of upper esophageal web was observed during the two-year follow-up.

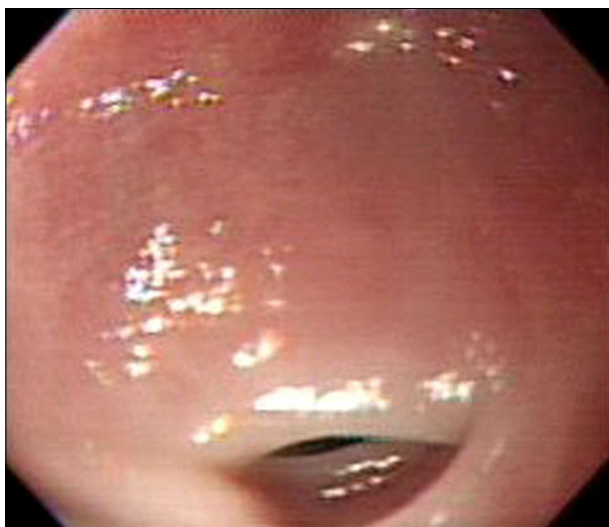


Fig. 5. Endoscopic findings when the web was relapsed. Relapse of esophageal web, which are similar to that observed previously.

DISCUSSION

The pathogenesis of web formation and dysphagia in Plummer-Vinson syndrome remains unclear⁵ and iron deficiency, genetic predisposition, or even autoimmune processes are considered as possible pathologies.⁴ Iron deficiency is hypothesized to reduce the levels of iron-dependent oxidative enzymes; this reduction gradually degrades pharynx muscles and cause atrophy of the mucosa, leading to the development of web.⁴ Another study showed that, low amplitude of contraction and high intrabolus pressure in the esophageal body was observed in a patient with Plummer-Vinson syndrome. After iron replacement, manometric examination of esophageal motility showed increased the amplitude of contraction and decreased intrabolus pressure.⁶

The diagnosis is based on the evidence of iron-deficiency anemia and one or more esophageal webs in a patient with postcricoid dysphagia. Esophageal webs can be detected by barium swallow X-ray and endoscopy. They appear smooth, thin, and gray with eccentric or central lumen. The webs typically occur in the proximal part of the esophagus.^{1,5}

Plummer-Vinson syndrome can be treated easily and effectively with iron supplementation and mechanical dilatation.¹ Iron replacement therapy may significantly improve dysphagia, especially in patients with web that are

not substantially obstructive.⁴ In case of significant obstruction of the esophageal lumen by esophageal web and persistent dysphagia despite iron supplementation, rupture and dilation of the web should be performed.¹ Obstructing webs yield to dilation and to cutting, either by using a needle-knife, papillotome, or argon plasma coagulation.⁷ Seo et al.⁷ reported the stenosis caused by the web was relieved by cutting the mucosal fold in 3 directions from the lumen to the wall, by using an insulated-tipped diathermic knife (IT2; Olympus). Enomoto et al.² reported successful endoscopic bougienage using a Celestin dilator (diameter, 12~18 mm). At the 3-year follow-up, their patient was healthy with no signs of relapse of anemia or dysphagia.² Katsinelos et al.⁸ reported recurrent esophageal web in Plummer-Vinson syndrome successfully treated with postdilatation intralesional injection of mitomycin C. But recurrent esophageal web has not been reported in Korea.

Surgical management is rarely necessary and the surgical indication and technique for the web have been controversial. Kitahara et al.⁹ reported surgical treatment case. Surgical removal of the web was attempted in October 1991 because of unsatisfactory results from conservative therapies and endoscopic bougienage. The operation was performed under general anesthesia by way of micro-laryngosurgery. The membranous structure was removed with a surgical knife and scissors through the inner lumen of esophagus. Prolene suture was used to close the incised mucosa. Esophagography, 6 months following the surgery revealed a well-dilated esophagus and no evidence of perforation. Endoscopic findings showed no recurrence of web 12 months post surgery.

Our patient was diagnosed with Plummer-Vinson syndrome at the first visit, for which she underwent endoscopic balloon dilatation because of near obstruction. After dilatation, dysphagia resolved. However, her anemic status remained uncorrected because she was lost to follow-up, and subsequently, esophageal web relapse occurred. Although the pathogenesis of esophageal web in Plummer-Vinson syndrome remains unclear, in light of our experience, iron-deficiency anemia has been implicated as the most important factor. We suggest the importance of continuous iron supplementation and long-

term follow-up in patients with Plummer-Vinson syndrome.

REFERENCES

1. Novacek G. Plummer-Vinson syndrome. *Orphanet J Rare Dis* 2006;1:36.
2. Enomoto M, Kohmoto M, Arafa UA, et al. Plummer-Vinson syndrome successfully treated by endoscopic dilatation. *J Gastroenterol Hepatol* 2007;22:2348-2351.
3. Sinha V, Prajapati B, George A, Gupta D. A case study of Plummer-Vinson syndrome. *Indian J Otolaryngol Head Neck Surg* 2006;58:391-392.
4. Atmatzidis K, Papaziogas B, Pavlidis T, Mirelis Ch, Papaziogas T. Plummer-Vinson syndrome. *Dis Esophagus* 2003;16:154-157.
5. Hoffman RM, Jaffe PE. Plummer-Vinson syndrome. A case report and literature review. *Arch Intern Med* 1995;155:2008-2011.
6. Dantas RO, Villanova MG. Esophageal motility impairment in Plummer-Vinson syndrome. Correction by iron treatment. *Dig Dis Sci* 1993;38:968-971.
7. Seo MH, Chun HJ, Jeon YT, et al. Esophageal web resolved by endoscopic incision in a patient with Plummer-Vinson syndrome. *Gastrointest Endosc* 2011;74:1142-1143.
8. Katsinelos P, Gkagkalis S, Chatzimavroudis G, Zavos C, Spyridakis J, Kountouras J. Recurrent esophageal web in Plummer-Vinson syndrome successfully treated with postdilation intraleisional injection of mitomycin C. *Gastrointest Endosc* 2012;75:1124.
9. Kitahara S, Ohmae Y, Ogura M, Matumura Y. The operation of upper esophageal web in Plummer-Vinson syndrome: a case report. *Auris Nasus Larynx* 1999;26:495-500.