



# Accidentally Confirmed Relapse of Plummer-Vinson Syndrome during Esophageal Foreign Body Evaluation

Hye Jin Kang, Seong Hun Kim, Seung Young Seo

Department of Internal Medicine, Jeonbuk National University Medical School, Research Institute of Clinical Medicine of Jeonbuk National University-Biomedical Research Institute of Jeonbuk National University Hospital, Jeonju, Korea

Plummer-Vinson syndrome (PVS) is a complex condition characterized by a classic triad of dysphagia, iron deficiency anemia (IDA), and an upper esophageal web. Dysphagia represents the most important symptom of PVS and is usually observed with solid food. Therapeutic options for dysphagia include iron supplementation and/or endoscopic balloon dilation based on symptom severity. PVS can be treated easily and regular iron replacement therapy can prevent disease progression. We describe a 63-year-old woman diagnosed with a simple esophageal web, who underwent esophageal dilation 13 years prior to presentation. However, she revisited our center because of dysphagia secondary to an esophageal foreign body, and endoscopy revealed esophageal web relapse. Investigations revealed IDA, and the patient was eventually diagnosed as PVS. She underwent successful endoscopic dilation and denied recurrent dysphagia after continuous intake of iron tablets. We recommend continuous management of IDA, as well as endoscopic dilation and a multidisciplinary therapeutic approach to ensure favorable prognosis in PVS patients. (**Korean J Helicobacter Up Gastrointest Res 2022;22:303-307**)

**Key Words:** Plummer-Vinson syndrome; Esophageal web; Iron supplementation; Balloon dilation

## INTRODUCTION

Plummer-Vinson syndrome (PVS) can be diagnosed following the discovery of a membranous web structure in the upper esophagus and iron-deficiency anemia (IDA) during an evaluation for dysphagia.<sup>1,4</sup> PVS, named after the doctor who first described it and also called Paterson-Kelly syndrome,<sup>5</sup> typically occurs in middle-aged women. Squamous cell carcinoma of the esophagus or pharynx occurs in 5.2~16% of patients, and gastric cancer may coexist in rare cases.<sup>3,4,6</sup> Although the prevalence and etiology of PVS have not been clearly identified, IDA is hypothesized to play the most important role in the formation of the upper esophageal web that causes the dysphagia.<sup>1,2,7</sup> Treatment is designed to relieve symptoms, and can be easily accomplished with iron supplementation alone or combined with endoscopic gastrointestinal dilatation, depending on the degree of dysphagia.<sup>1,2</sup> The prog-

nosis is good, and recurrence is rare when iron supplementation is continued.<sup>8</sup>

A middle-aged woman visited our center because of dysphagia, and was diagnosed with a simple esophageal membrane, despite endoscopic evidence of an upper esophageal web and blood tests that revealed IDA. She underwent digestive tract dilatation and received no further follow-up. Subsequently, she developed dysphagia due to recurrence of the esophageal membrane, and PVS was diagnosed. Herein, we describe a PVS patient who was treated at our center (IRB number: CUH 2022-08-033).

## CASE REPORT

A 63-year-old woman visited the emergency room with dysphagia that began after dinner on the day prior to admission. The patient was unable to even swallow water, despite reporting no previous problems eating solid food. Her only significant medical history was an esophageal stricture dilatation, 13 years earlier, when she had been diagnosed with an upper esophageal web. The diagnosis and treatment were subsequent to an emergency room visit due to dysphagia that had progressively worsened

Received: August 5, 2022 Revised: August 22, 2022 Accepted: September 1, 2022

Corresponding author: Seung Young Seo

Department of Internal Medicine, Jeonbuk National University Medical School, Research Institute of Clinical Medicine of Jeonbuk National University-Biomedical Research Institute of Jeonbuk National University Hospital, 20 Geonji-ro, Deokjin-gu, Jeonju 54907, Korea

Tel: +82-63-250-2676, Fax: +82-63-254-1609, E-mail: bear7905@jbnu.ac.kr

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over a 4-year period (Fig. 1). There were no significant findings in her medication and family history.

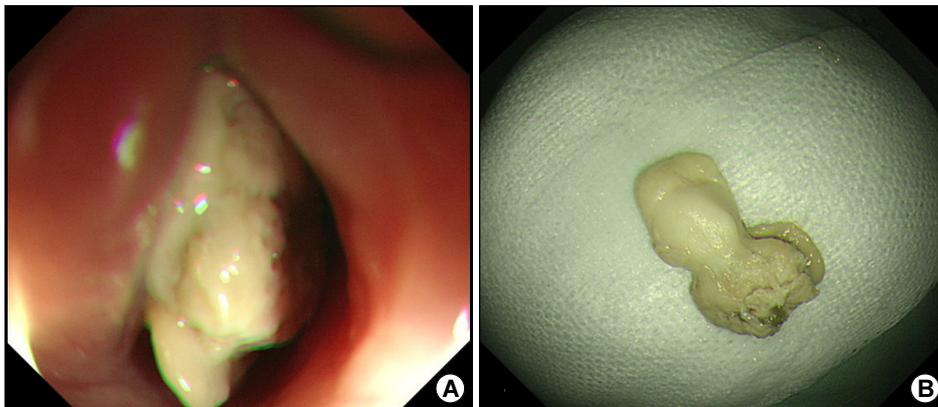
A physical examination at the time of admission revealed a blood pressure of 114/63 mmHg, pulse rate of 80 beats/min, respiratory rate of 20 beats/min, and a body temperature of 36.8°C. The patient's general condition was good, although her conjunctiva were slightly pale. No other specific physical findings, such as oral cavity glossitis, were observed.

At the time of admission, the leukocyte count was 2,670/mm<sup>3</sup> (index, 4,800~10,800/mm<sup>3</sup>), hemoglobin level was 9.7 g/dL (index, 12~16), hematocrit was 30.5% (index,

37~48%), and platelet count was 185,000/mm<sup>3</sup> (index, 130,000~450,000/mm<sup>3</sup>). The mean hematocrit, mean amount of erythrocyte pigment, and mean erythrocyte pigment concentration were reduced to 76.9 fL (index, 81~99), 22.6 pg (index, 27~32) and 31.8 g/dL (index, 33~37), respectively. The patient's serum iron concentration and transferrin saturation were also decreased to 14 µg/dL (index, 53~167) and 3.4% (index, 20~50%), respectively. Her serum ferritin level was 15.8 ng/mL (index, 10~291), which was within the lower limit of normal level. Moreover, her folic acid (6.89 ng/mL [index, 1.5~16.9]) and vitamin B12 (180.5 pg/mL [index, 160~970]) levels were within normal ranges. A peripheral blood smear showed microcytic hypochromic anemia, suggesting IDA. The peripheral blood smear taken during the emergency room visit 13 years earlier also showed findings consistent with IDA. The upper gastrointestinal endoscopy show a foreign body in the narrowed part of the upper esophagus. The foreign body in the esophagus was removed by using a forceps and was identified as a clam (Fig. 2). After the foreign body removal, endoscopy was performed again. A transparent, pearly, thin membrane structure was observed in the lower part of the upper esophageal sphincter, 15 cm from the incisors. The remaining space in the esophagus was too narrow even for an endoscope for infants (Fig. 3). PVS was diagnosed based on the dysphagia, IDA, and upper esophageal findings. Endoscopic balloon dilatation was performed because oral food intake was deemed difficult due to the



**Fig. 1.** Endoscopic image of the esophagus taken 13 years prior to presentation. An upper esophageal web is observed.



**Fig. 2.** Endoscopic findings of an upper esophageal foreign body and after the removal. (A) Foreign body impaction is visualized at the upper esophagus. (B) Image showing removal of a Manila clam using tripod grasping forceps.

narrowed esophageal lumen (Fig. 4).

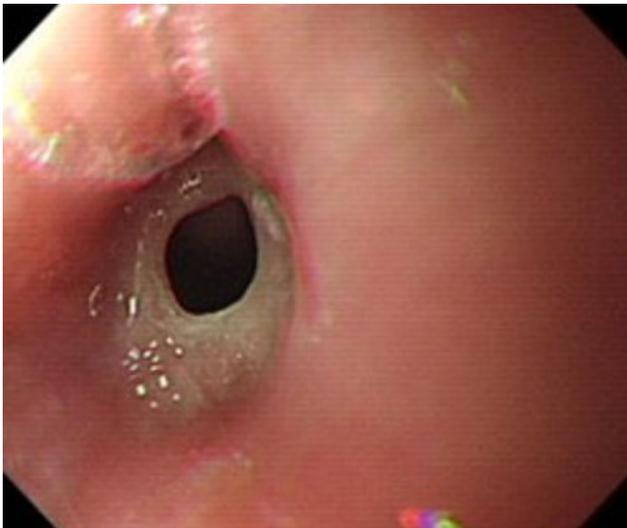
Subsequently, the patient was able to swallow solid foods, and she was discharged with a prescription for iron supplement. During a colonoscopy, there was no abnormal finding related to IDA.

## DISCUSSION

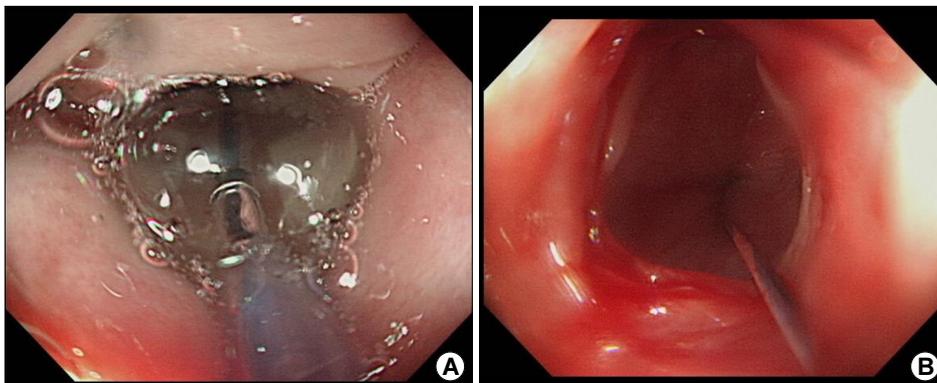
An esophageal web is a thin (<2 mm thick) mucous membrane that is smooth and gray in color. It is often located below the cricoid cartilage of the upper esophagus, protruding into the esophageal lumen and blocking part of the lumen.<sup>1,6</sup> The incidence is about 1% of the total

population and is found in 5~15% of dysphagia patients.<sup>9</sup> PVS is a well-known cause of dysphagia.<sup>10</sup> Most PVS patients are initially asymptomatic. As it progresses over several months to years, dysphagia starts to occur, especially when eating solid foods.<sup>1,2,10,11</sup> Barium esophageal angiography, video fluoroscopy, and endoscopy are useful tests for diagnosing the esophageal web.<sup>1,12</sup>

Another typical clinical symptom of PVS is IDA.<sup>1,2</sup> Although the cause of the esophageal web is not yet clearly identified, IDA is accepted as the most valid cause among the several hypotheses including malnutrition, hereditary factors, autoimmune factors, and vitamin deficiency.<sup>1,2,7</sup> When iron deficiency exists, the activity of iron-dependent oxidative enzymes decreases and atrophy occurs in the tongue, pharynx, and gastric mucosa. The atrophied mucous membrane, especially that in the pharynx, is prone to inflammation. Eventually fibrosis is induced in the surrounding muscles, causing their gradual degeneration. Specifically, the mucous membrane under the cricoid cartilage is where maximum mechanical stimulation occurs while swallowing solid foods, leading to accumulated trauma. The iron deficiency hypothesis suggests that the inflammatory response induced by the atrophy and the damage to the mucous membrane contributes to the formation of the esophageal web, thereby causing dysphagia.<sup>7,12</sup> This hypothesis does not explain why the degree of anemia and the dysphagia symptoms are not proportional, why the esophageal web is limited to the upper esophagus, and why some esophageal webs develop even in the absence of anemia. However, it is evident



**Fig. 3.** Endoscopic image after foreign body removal. An esophageal web is visualized 15 cm from the incisors.



**Fig. 4.** Endoscopic findings showing endoscopic balloon dilation for management of the esophageal web. (A) Image showing the endoscopic balloon dilation procedure. (B) The web is dilated after the balloon dilation.

that IDA is closely related to the diagnosis and treatment of PVS because dysphagia is relieved by correcting only the iron deficiency.<sup>7,10</sup>

Treatment of PVS primarily involves the non-invasive administration of iron; however, if the symptoms do not improve after iron intake or if iron supplements cannot be administered, invasive methods (e.g., endoscopic swelling or balloon dilatation, electrocautery, and, rarely, surgery) may be considered.<sup>1,3,8,9</sup> Importantly, prior to beginning iron supplements to correct the IDA, the cause of the anemia must be determined and appropriately treated. Iron deficiency is most often caused by excessive menstruation or chronic blood loss from the gastrointestinal tract including peptic ulcers, hemorrhoids, vascular abnormalities, and malignant diseases. In addition, partial gastrectomies, uterine fibroids, and chronic inflammatory diseases have been reported as causes.<sup>10,13</sup> Another caution is that iron supplementation needs to be maintained. As in the present case, Won et al.<sup>8</sup> reported dysphagia recurrence because the IDA was not corrected after endoscopic balloon dilatation.

Previous studies have reported that upper gastrointestinal tract squamous cell carcinoma occurs in about 3~16% of patients with PVS. Further, esophageal webs are considered precancerous lesions of the upper esophageal tumors. In a recent single-center study in India, cancer was found in the hypopharynx and central esophagus in six out of 132 patients (4.5%), with rare cases accompanying advanced gastric cancer.<sup>4,6</sup> In addition to preventing the recurrence of symptoms, continuous iron supplementation is required until the anemia is corrected in order to prevent the progression of the esophageal web to oral, pharyngeal, esophageal, and/or gastric cancer.<sup>3,4,6,10</sup>

Post-treatment outcomes are good, and patients usually require only a single endoscopic dilatation to improve dysphagia. If iron supplementation is continued, subsequent recurrence is rare. As with most IDA patients, iron supplementation typically involves oral administration of ferrous sulfate (150~200 mg/day based on the dose of elemental iron, divided into 2~3 split doses taken on an empty stomach).<sup>14</sup> In general, hemoglobin levels are restored after taking iron supplements for about 6~8

weeks.<sup>14</sup> To accumulate sufficient stored iron, an additional dose is recommended for next 6-12 months, along with periodic checks to determine whether the IDA has recurred. In addition, since malignant tumors of the upper gastrointestinal tract may occur identification and monitoring are important. PVS is a complex disease and the involvement of a multidisciplinary integrated treatment is expected. The diagnosis, treatment, and follow up of PVS should involve specialists in gastroenterology, hematology, oncology, radiology, and surgery.<sup>1,4,12</sup>

In the present case, the patient was previously diagnosed as having a simple esophageal membrane 13 years before her admission; thereafter, she was discharged after receiving only gastrointestinal dilatation. At that time, the patient visited the hospital with a 4-year history of progressive dysphagia. Nonetheless, this is a typical case of PVS in which an esophageal web and IDA were confirmed. The esophageal membrane recurred as the patient did not receive adequate iron supplementation. Regardless, the disease recurrence was initially identified as acute dysphagia due to a foreign body, but the patient was eventually diagnosed, 17 years after the initial symptom. During the present admission, dysphagia and test findings showed improvement after endoscopic esophageal dilatation and iron supplementation. Periodic examinations will be conducted with cancer surveillance to monitor the esophageal web and anemia. This case emphasizes the importance of maintaining appropriate iron supplementation to prevent PVS recurrence, in addition to mechanical dilation for the dysphagia treatment. Further, this case highlights the need for a multidisciplinary approach and emphasizes the need for making an accurate diagnosis when an esophageal web is observed in a patient who complains of dysphagia.

## CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

## SUPPLEMENTARY MATERIAL

Supplementary material 1. Korean translation of the

articles available from <https://doi.org/10.7704/kjhugr.2022.0036>.

## ORCID

Hye Jin Kang  <https://orcid.org/0000-0001-9186-0791>  
 Seong Hun Kim  <https://orcid.org/0000-0002-7592-8060>  
 Seung Young Seo  <https://orcid.org/0000-0003-2018-0013>

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