

# A Rare Cause of Plummer-Vinson Syndrome: Celiac Disease

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## Abstract

Plummer-Vinson syndrome (PVS) is a clinical condition accompanied by dysphagia, iron deficiency anemia and esophageal web. PVS has not been encountered in recent years since the factors related to nutritional have been improved and iron deficiency anemia has been diagnosed and treated early. A rare cause of PVS is Celiac disease. There are very few cases reported in the literature related to Celiac disease and PVS coexistence. We wanted to present this case both to emphasize this togetherness PVS is rarely seen and because PVS may still be seen today.

Keywords: Celiac disease; iron deficiency anemia; plummer-Vinson syndrome.

Plummer-Vinson syndrome (PVS) is a condition characterized by dysphagia, iron deficiency anemia and upper esophageal web. PVS was first described by Plummer in 1912 with a series of 21 cases [1]. Seven years later (in 1919), new cases with similar characteristics were attributed to these cases, which had been previously reported by Vinson [2]. The etiopathogenesis of PVS is not known precisely. It is discussed in the literature that iron deficiency and autoimmune causes pave the way for web formation in genetically predisposed individuals and also a reduction in oxidative enzymes due to iron deficiency causes atrophy in pharyngeal muscles and esophageal mucosa, which also leads to web formation. This theory is also supported by clinical reports of dysphagia improvement after iron ther-

apy [3]. It is seen in PVS that the squamous cell carcinoma of the esophagus in the first place, especially the esophagus, pharynx and other cancers of the gastrointestinal tract, were found to increase by 3-15% as well. Thus, annual endoscopic follow-up is recommended to patients with PVS [4,5]. PVS may arise from many clinical conditions that lead to iron deficiency. Celiac disease is a rare cause of PVS. There are very few cases reported in the literature regarding the coexistence of PVS and Celiac disease [6,7]. A case in which Celiac disease, PVS and post cricoid carcinoma were seen together was also been reported [8]. We should note that it is not known how Celiac disease causes PVS development. PVS is not expected to occur in developed societies because of the early diagnosis and treatment of iron deficiency in recent

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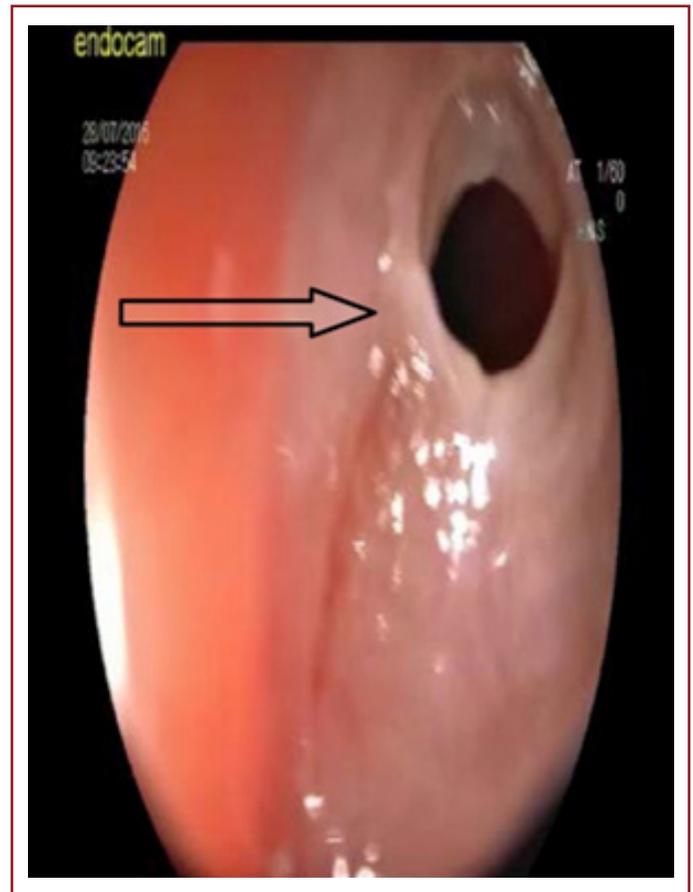
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years and the partly correction of nutritional factors globally. However, PVS may still be seen in socioeconomically low living space and developing countries. In this case, PVS and Celiac disease were diagnosed in a young woman who applied with symptoms of dysphagia and anemia, and because of this togetherness that is seen rarely, we considered this study appropriate to share with you.

## Case Report

Patient's consent was obtained for this study. An 18-year-old woman applied to the gastroenterology polyclinic because of ever-increasing complaints of dysphagia, dizziness and weakness for the last two years. There was no significant feature in the patient's backstory and family history. In her physical examination, it was revealed that her conjunctival was pale and she had tachycardia. The trachea was in close, and her spleen was palpable for 2-3 cm in lower of costae level. Her values in accordance with laboratory examination were haemoglobin: 6.6 g/dL (12.9-18.1), platelet count: 375 thousand/uL (142-424), MCV: 53 fL (80-97), RDW: 15% (11.6-15.8), iron: 9 ug/dl (47-169), iron-binding capacity: 489 ug/dl (155-300) and ferritin: 2 ng/dL (11-306.8). Peripheral smear was compatible with the hypochromic microcytic anemia. The patient did not describe melena, rectal bleeding or excessive menstrual bleeding. There was no obvious diarrhea. Vitamin B12, thyroid function tests, vitamin D and folic acid levels were normal. Antiendomysium and tissue transglutaminase antibodies were examined in the patient because she might have iron deficiency anemia due to Celiac disease. These tests were found to be highly positive. An upper gastrointestinal endoscopy was planned to confirm the diagnosis of Celiac disease and to investigate the etiology of dysphagia. In the endoscopy, it was observed the web that narrowed the lumen as partially in just below the upper esophageal sphincter (Fig. 1). This place was passed with an endoscope pushing lightly, which caused the web rupture. However, there was no significant bleeding and the examination continued. The esophagus and stomach were normal. The bulb and the second part of the duodenum were in nodular, combed appearance. A large number of biopsies were taken from these sections. Biopsy results were reported consistent with Celiac disease involving total villus atrophy and crypt hyperplasia. Thus, Celiac disease was diagnosed. It was reached the conclusion that the Celiac disease might arise from a lack of iron and then PVS. It was begun to be applied parenteral iron replacement to the patient, and she was followed by suggesting a gluten-free diet. Dysphagia improved due to the rupture of the web-based on the endoscopic procedure



**Figure 1.** Esophageal web.

performed three months later and by the help of iron treatment. It was observed that Hgb increased to 10 gr and the iron parameters were significantly improved.

## Discussion

PVS is characterized by a triad of dysphagia, anemia of iron deficiency and esophageal web. Many clinical conditions that cause iron deficiency may lead to PVS. Although PVS is usually seen in middle-aged patients, it can also be seen in child and adolescent periods [9,10]. PVS is closely related to pharynx and cancers of the digestive tract. Thus, endoscopic follow-up is recommended [4,5]. PVS is seen as more common in low socioeconomic regions where poor nutritional conditions and nutritional deficiencies are present. The togetherness of Celiac disease and PVS is a very rare condition. The webs seen in PVS are usually in the anterior wall of the esophagus and are crescent-shaped, rarely concentric. The webs are usually in thin membranous appearance and covered with squamous epithelium. The most important diagnostic methods for showing the esophageal webs are barium esophagus graphy and upper gastrointestinal endoscopy [11,12]. Symptoms improve with iron

replacement in a fraction of patients who have dysphagia affiliating to the Web. The mechanical methods, such as bougie, balloon dilatation or endoscopic surgical methods, such as incision with electrocautery, can be applied to patients whose dysphagia continues despite the iron replacement [13–15]. Our patient was an 18-year-old woman who applied to our clinic with complaints of dysphagia and fatigue, which progressively increased over the last 2-3 years. Deep iron deficiency anemia was detected in the patient's examination. Iron deficiency anemia was found to be related to Celiac disease. Upper gastrointestinal system examination was performed to investigate dysphagia etiology and to confirm Celiac disease. On the endoscopic examination, an appearance fitting to the thin web with a membranous character was detected just below the upper esophageal sphincter. When trying to pass to distale, the web was ruptured, but it was continued to be investigated as there was no obvious bleeding. In the examination of the duodenum, it was seen that the mucosa was atrophic and nodular in appearance. The mucosa was taken biopsies for Celiac disease. When biopsies and serologic tests for Celiac disease were detected as positive, Celiac disease was diagnosed. Her dysphagia was symptomatic due to the rupture of the web after the endoscopic procedure was resolved. It was begun to be applied a gluten-free diet and parenteral iron treatment to the patient. In the controls, there was a significant improvement in the parameters examined for anemia of iron deficiency. Endoscopic follow-up was also recommended concerning possible malignancies that could be developed.

**Informed Consent:** Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

**Peer-review:** Externally peer-reviewed.

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