Turk J Gastroenterol *2016; 27: 81-2*

Two cases with Plummer–Vinson syndrome in the 21st century

İlhami Gültepe[1], Metin Başaranoğlu[2]

[1]Department of Internal Medicine, Bezmialem Vakıf University Hospital, İstanbul, Turkey
[2]Department of Internal Medicine, Division Gastroenterology, Bezmialem Vakıf University Hospital, İstanbul, Turkey

ABSTRACT

Plummer–Vinson syndrome (PVS) is characterized by the presence of postcricoid dysphagia, iron deficiency anemia, and upper esophageal web. This syndrome is now a rare condition because of the improvement in nutritional status and increased awareness regarding iron deficiency anemia and the early diagnosis and easy treatment of this anemia or underlying causes. In this presentation, we report two middle-aged female patients with PVS and briefly review the literature.

Keywords: Dysphagia, anemia, upper esophageal web, Plummer–Vinson syndrome

Case Report

INTRODUCTION

The Plummer–Vinson syndrome (PVS) is characterized by the presence of postcricoid dysphagia, iron deficiency anemia, and upper esophageal web (1,2). This syndrome is a rare condition in the 21st century because of the improvement in nutritional status, advancement in medical tools, and increased awareness about iron deficiency anemia and the early diagnosis and treatment of this condition or underlying causes. In this presentation, we report two middle-aged female patients with upper esophageal web who presented with dysphagia in the clinical practice. Further evaluations showed that these two patients had PVS. We also briefly reviewed the PVS literature.

CASE PRESENTATIONS

Case 1

A 45-year-old woman presented with abdominal pain, loss of appetite, difficulty in swallowing solids and liquids, cough, and sputum. She had progressive dysphagia for the last 10 years. She did not have any other disease or drug or alcohol consumption or smoking habit. Her physical examination revealed anemia, glossitis, and cheilitis. Laboratory evaluations revealed a hemoglobin level of 9.6 g/dL, white blood cell count of 5.700/mm, platelet count of 453.000/mm, iron concentration of 12 µg/dL (normal concentration: >21 µg/dL for female), and ferritin concentration of 5.7 ng/mL (normal concentration: 20–495 ng/mL for females). A barium study of the esophagus showed a narrow lumen of the upper esophagus (Figure 1). Upper gastrointestinal endoscopy showed a typical circumferential membranous web on the upper esophagus. The rest of the upper gastrointestinal tract could not be observed because of the web. Biopsies were performed endoscopically from the web, and the histological examination of these specimens showed plasma cells, lymphocytes, and histiocytic infiltration. Parenteral iron administration and close follow-up were recommended. The patient was examined by radiological techniques 2 months later, and the lumen was expanded meaningfully.

Case 2

A 45-year-old woman complained of difficulty in swallowing solids for the last 6 years. The nature of the dysphagia was progressive. She did not have any other disease or drug or alcohol consumption or smoking habit. She reported having eaten soil 20 years ago for a short period. A barium swallow test revealed narrowing of the area at the proximal part of the upper esophagus. Laboratory evaluations revealed a hemoglobin level of 10.6 g/dL, white blood cell count of 6.400/mm, platelet count of 405.000/mm, and iron concentration of 17 µg/dL (normal concentration: >21 µg/dL for females). A tiny mucosal membrane of the upper esophagus was

Address for Correspondence: Metin Başaranoğlu E-mail: basaranoglu@hotmail.com metin_basaranoglu@yahoo.com
Received: October 23, 2015 **Accepted:** October 28, 2015 **Available Online Date:** November 27, 2015

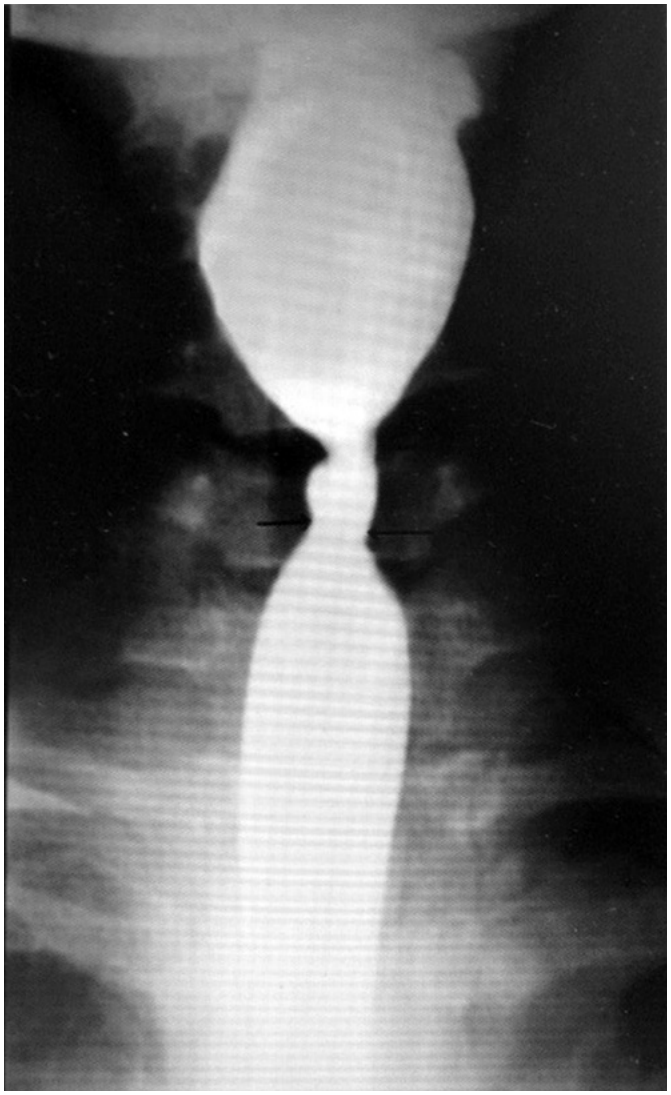

Figure 1. Barium study of the esophagus showing upper esophageal web.

observed, and it was ruptured by the endoscope. The rest of the upper gastrointestinal tract was normal. Oral iron administration was recommended. The patient was also recommended for bougienage of the membrane. Both female patients in this presentation were diagnosed on the basis of the presence of dysphagia, iron deficiency anemia, and upper esophageal webs. Dysphagia was due to the web formation in both cases, and the webs were smooth, tiny, and gray.

DISCUSSION

PVS is a disease most commonly found in middle-aged females. Currently, it is a rare condition because of better nutrition and healthcare conditions (1,2). Most webs are located along the anterior esophageal wall in the shape of a crescent or less frequently concentric. This tiny mucosal membrane is covered by normal squamous epithelium. Barium esophagram and upper gastrointestinal endoscopy are the preferred diagnostic tests used to detect esophageal webs (1,2). Esophageal or pharyngeal cancer can develop in 3%–15% of patients with PVS. Thus, yearly surveillance endoscopy is recommended for patients with PVS (3,4). Adult celiac disease might present as refractory anemia. Thus, there have been reported cases of celiac disease associated with PVS due to refractory iron deficiency anemia (3,4). Therefore, random duodenal biopsies in all patients with PVS were recommended. We did not perform biopsy in our cases because we found a normal duodenal mucosal pattern during the endoscopy in the second case. Although we failed to reach the second segment of the duodenum in the first case, her dysphagia was resolved by iron replacement therapy, and the narrow lumen was expanded meaningfully using barium radiography. These improvements excluded refractory anemia due to celiac disease in this case as well. Upper gastrointestinal endoscopy and forceful dilatation or rupture of the web are generally sufficient to improve the swallowing difficulty and relieve dysphagia in most of the patients (5,6). Dysphagia associated with PVS also improved with the administration of iron. The exact cause and pathophysiology of PVS development remain unclear. Proposed mechanisms include iron and other nutritional deficiencies, genetic predisposition, and autoimmunity (1,2). It is considered that the disturbance of muscles due to the deficiency of iron-dependent oxidative enzymes, esophageal mucosal atrophy, and epithelial disturbance causes esophageal webs. In conclusion, we would like to highlight that PVS should still be considered in the differential diagnosis of dysphagia in the 21st century.

Ethics Committee Approval: N/A.

Informed Consent: Written informed consent was obtained from patients who participated in this case.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - I.G., M.B.; Design - I.G., M.B.; Supervision - I.G., M.B.; Resource - I.G., M.B.; Materials - I.G., M.B.; Data Collection and/or Processing - I.G., M.B.; Analysis and/or Interpretation - I.G., M.B.; Literature Search - I.G., M.B.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

REFERENCES

1. Atmatzidis K, Papaziogas B, Pavlidis T, Mirelis Ch, Papaziogas T. Plummer-Vinson syndrome. Dis Esophagus 2003; 16: 154-7. [CrossRef]
2. Geerlings SE, Statius van Eps LW. Pathogenesis and consequences of Plummer-Vinson syndrome. Clin Investig 1992; 70: 629-30. [CrossRef]
3. Rashid Z, Kumar A, Komar M. Plummer-Vinson syndrome and postcricoid carcinoma late complications of unrecognized celiac disease. Am J Gastroenterol 1999; 94: 1991. [CrossRef]
4. Jessner W, Vogelsang H, Puspok A, et al. Plummer-Vinson syndrome associated with celiac disease and complicated by postcricoid carcinoma and carcinoma of the tongue. Am J Gastroenterol 2003; 98: 1208-9. [CrossRef]
5. Huynh PT, de Lange EE, Shaffer HA. Symptomatic webs of the upper esophagus: treatment with fluoroscopically guided balloon dilation. Radiology 1995; 196:789-92. [CrossRef]
6. Bredenkamp JK, Castro DJ, Mickel RA. Importance of iron repletion in the management of Plummer-Vinson syndrome. Ann Otol Rhinol Laryngol 1990; 99: 51-4.

Case Report