Case Report

Bagcilar Medical Bulletin 2017;2(1):25-28
DOI: 10.5350/BMB20161224025457

Endoscopic Management of Dysphagia in Plummer–Vinson syndrome

Ali Solmaz[1], Osman Bilgin Gulcicek[1], Talar Vartanoglu[1], Erkan Yavuz[1], Hakan Yigitbas[1], Candas Ercetin[1], Atilla Celik[1], Fatih Celebi[1]

[1]Bagcilar Training and Research Hospital, General Surgery Clinic, Istanbul-Turkey

Corresponding author:
Ali Solmaz,
Bagcilar Training and Research Hospital, General Surgery Clinic,
Merkez Mahallesi, Mimar Sinan Caddesi, 6. Sokak 34200 Bagcilar/Istanbul, Turkey
Phone: +90-212-440-4000/2081
Fax: +90-212-440-4242
E-mail address: solmazali@hotmail.com

Date of submission: November 24, 2016
Date of acceptance: December 24, 2016

ABSTRACT

Plummer-Vinson syndrome (PVS), (also called Paterson-Brown-Kelly syndrome or sideropenic dysphagia) is an extremely rare syndrome which includes iron-deficiency anemia, dysphagia, and esophageal webs. If left untreated, progressive dysphagia can supervene and the patient is at risk of developing pharyngeal or esophageal squamous-cell carcinoma. Treatment consists of supplementation of iron and endoscopic dilations or surgery for the esophageal web. We describe the case of a 17-year-old women with Plummer-Vinson syndrome whose dysphagia was successfully treated with endoscopic dilation. She remains in good general condition 3 months after treatment.

Keywords: dysphagia, esophageal web, iron-deficiency anemia, Plummer-Vinson syndrome

ÖZET

Plummer-Vinson sendromunda yutma güçlüğünün endoskopik tedavisi

Plummer-Vinson sendromu (PVS), (Peterson-Brown-Kelly sendromu veya sideropenik disfaji olarak da bilinir) oldukça nadir görülen bir sendrom olup demir eksikliği anemisi, özefagusta web oluşumu ve buna bağlı yutma güçlüğü ile karakterizedir. Tedavi edilmediği takdirde yutma güçlüğü giderek artar ve bu hastalar uzun vadede özefagus ve farenksin skuamöz hücreli kanserinin riski altındadırlar. Tedavi demir replasmanı ve özefagustaki webin endoskopik veya cerrahi olarak açılmasından ibarettir. Biz burada kliniğimize yutma güçlüğü ile başvuran ve endoskopik balon dilatasyonu ile başarılı bir şekilde komplikasyonsuz olarak yutma güçlüğü tedavi edilen 17 yaşındaki olgumuzu sunmak istedik.

Anahtar kelimeler: yutma güçlüğü, özefajeal web, demir eksikliği anemisi, Plummer-Vinson sendromu

Introduction

Plummer-Vinson syndrome or Paterson-Kelly syndrome (PVS) is defined by the classical triad of iron-deficiency anemia, dysphagia, and esophageal webs. The name of the syndrome refers to two physicians of the Mayo Clinic, Henry Stanley Plummer and Porter Paisley Vinson (1). Another name of the condition, Paterson-Kelly syndrome, goes back to two British laryngologists (Donald Ross Paterson and Adam Brown-Kelly) who first described its characteristic clinical features (2,3). Patients' main complaint is gradually progressing difficulty in swallowing solid foods. At the same time, the patient may have atrophic oral mucosa, cracks or fissuring at the corners of the mouth, glossitis, koilonychia, or nails that are brittle and break easily (4). Over the years, dysphagia gradually progresses, and the patient increasingly finds it difficult to swallow solid food. Therapeutic options of PVS consist of dilation and surgery. Although surgery tends to be reserved for ineffective dilation, efficacy and risk of dilation have been controversial (5). As a technique of dilation, there are dilators or balloon dilator; however, there has been no comparison study of these. The balloon dilator may be the preferred instrument because its expanding force can be focused on the stenotic segment without shear stress (6,7). This approach is more efficient and secure.

In this report, we present a young female patient with upper esophageal web, who presented with long-standing dysphagia and sideropenia and was treated effectively with endoscopic balloon-dilation therapy and iron supplementation.

Case Report

A 17-year-old woman presented with difficulty in swallowing solid food for 10 months. She did not display any other disease or drug or alcohol consumption or smoking habit. Her physical examination revealed anemia, glossitis, and cheilitis. Laboratory investigations revealed a hypochromic microcytic anemia with a hemoglobin level of 6.9 g/dL (normal range is 11.3-13.4), mean corpuscular volume (MCV) of 63.34 fl (normal range is 80-99), iron concentration of 15 µg/dL (normal concentration: >30 µg/dL for females), and ferritin concentration of 12.9 ng/mL (normal concentration: 13-150 ng/mL for females). The patient's esophagogram showed the presence of esophageal web (Figure 1). Upper gastrointestinal endoscopy showed a typical circumferential membranous web in the area of the upper esophagus. The remainder of the upper gastrointestinal tract could not be evaluated because of the web (Figure 2). Fulfilling the classical triad of dysphagia, anemia, and upper esophageal web, Plummer-Vinson syndrome was diagnosed.

Enteral iron administration (150 mg daily) was recommended immediately with close follow-up. Endoscopic balloon dilation was carried out successfully with the assistance of guidewire by sight (Figure 2). After balloon dilation, the endoscope passed through the esophagus easily and the remainder of the upper gastrointestinal tract was examined; the rest of the esophagus, stomach, and duodenum were macroscopically normal. The patient has been continuing the intake of iron (III) supplement for three months and was found to be in good general condition; there is no symptom of difficulty in swallowing.

Discussion

The predominant classical triad of the Plummer-Vinson or Paterson-Kelly syndrome consists of upper esophageal web or webs, dysphagia, and iron-deficiency anemia. Data for incidence and prevalence of the syndrome are not available. It seems to have been more common in Caucasians of Northern countries, particularly among middle-aged women (8). However, it is extremely rare nowadays. The decline in reports and incidence of the syndrome may be related to the improvement in nutritional status and better treatment of iron deficiency (9). In Turkey, however, many Plummer-Vinson syndrome cases are still being reported (10,13,14).

The most common benign cause of the cervical type of dysphagia is an upper esophageal web. These webs may be found in approximately 5-15% of patients with dysphagia. Webs are fragile membranes and respond well to esophageal dilation therapy (11). This minute mucosal membrane is covered by normal squamous epithelium. The exact cause and pathophysiology of PVS development remain unclear. It is considered that the disturbance of muscles due to the deficiency of iron-dependent oxidative enzymes, esophageal mucosal atrophy, and epithelial disorder may cause esophageal webs.

Barium esophagogram and upper gastrointestinal endoscopy are the preferred diagnostic tests used to detect esophageal webs (4,12). Upper gastrointestinal endoscopy and forceful dilation or rupture of the web are generally sufficient to improve the swallowing difficulty and relieve dysphagia in

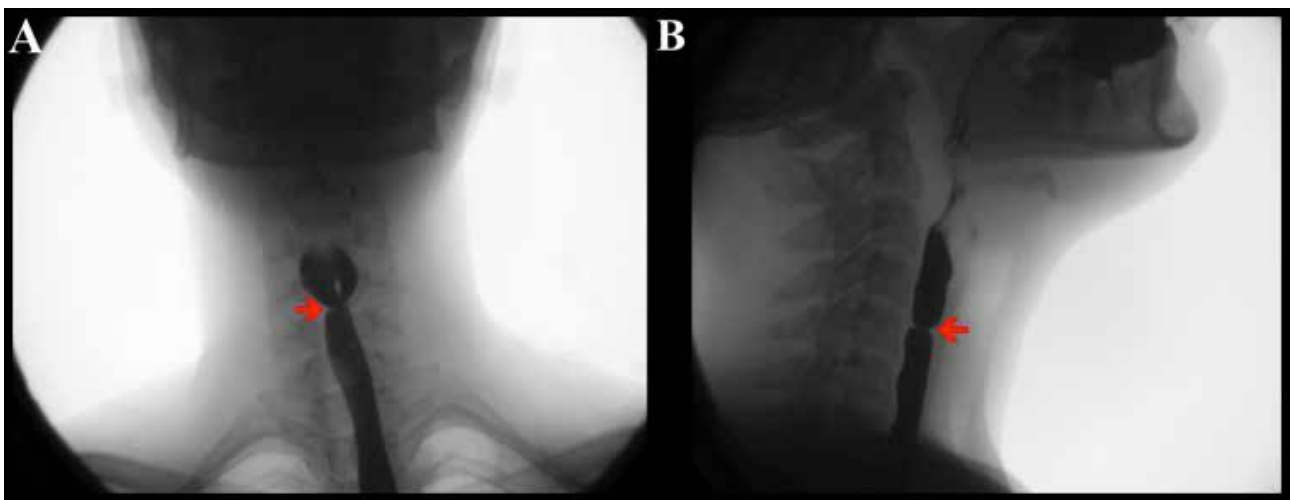

Figure 1: Radiographic study of the patient showing indentation (red arrows) of the proximal esophagus indicating esophageal web. (A: Anteroposterior view, B: Lateral view)

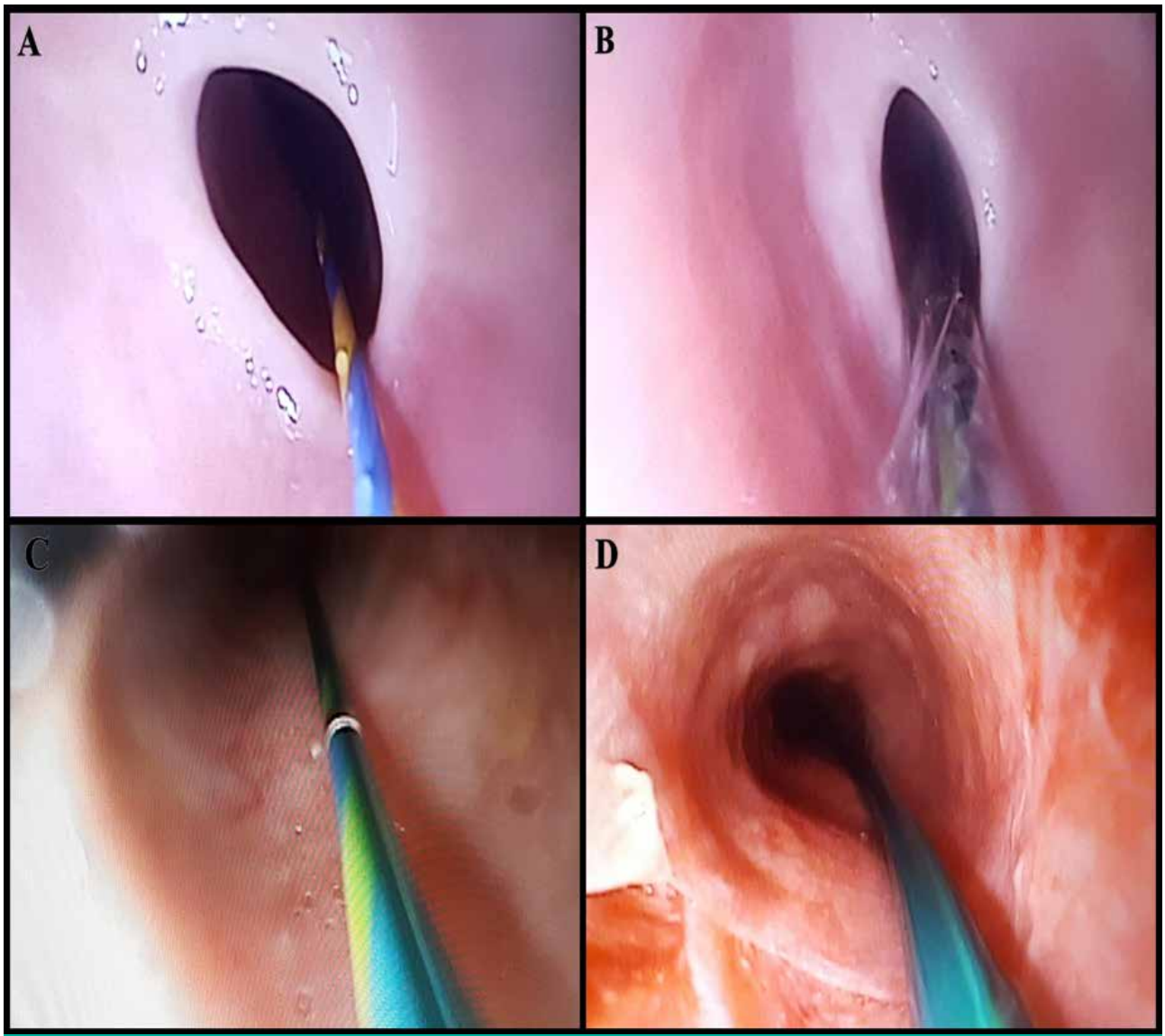

Figure 2: Endoscopic view of esophageal web dilation. (A: Esophageal web, B: Balloon is inserted through the guidewire, C: Image after dilation of transparent balloon, D: After dilation of web.)

most of the patients (13). In our case, dysphagia was the main symptom. Plummer-Vinson syndrome affects mainly white women in the fourth to seventh decade of life, but some cases in children and adolescents have been reported (14,15). The pathogenesis of the syndrome remains unclear; possible etiopathogenetic mechanisms include iron deficiency, genetic predisposition, or autoimmune disorder. It is reported that iron deficiency leads to the reduction of iron-dependent oxidative enzymes, which results in gradual degradation of muscles of the pharynx. In turn, mucosal atrophy may cause the development of webs. Plummer-Vinson syndrome can be accompanied by pernicious anemia, thyroiditis, or celiac disease (16).

The treatment of Plummer-Vinson syndrome is iron supplementation, and iron therapy may be necessary in the presence of a web formation even though hematologic parameters are normal. Patients with dysphagia, choking, and episodes of aspiration need dilation therapy together with iron supplementation. Endoscopic dilation is simple and a preferred procedure in the treatment of the syndrome and of cervical web of the esophagus (11). It is important to differentiate this syndrome from other causes of dysphagia, e.g. malignant

tumors, strictures, esophageal burns, heterotopic gastric mucosa, or blistering skin disease. Plummer-Vinson syndrome is known to be associated with upper alimentary tract cancer (squamous cell carcinoma), and surveillance endoscopy is recommended (17,18). In our patient, treatment was achieved by dilation and iron therapy. Although bougienage dilation was preferred in some previous reports (13), we experienced that balloon dilation was efficient and reliable, because we dilate the web with the help of guidewire by sight. Finally, we would like to highlight that PVS should still be considered in the differential diagnosis of dysphagia.

Contribution Categories	Name of Author
Follow up of the case	A.S., O.B.G., E.Y., T.V., H.Y., C.E., F.C.
Literature review	A.S., O.B.G., E.Y., H.Y., C.E., A.C.
Manuscript writing	A.S., O.B.G., T.V., C.E., F.C., A.C.
Manuscript review and revisation	A.S., E.Y., T.V., H.Y., A.C., F.C.

Conflict of Interest: Author declared no conflict of interest.

Financial Disclosure: Authors declared no financial support.

References

1. Novacek G. Plummer-Vinson syndrome. Orphanet J Rare Dis 2006;1:36. [CrossRef]
2. Kelly AB. Spasm at the entrance of the esophagus. J Laryngol Otol 1919;34:285-289. [CrossRef]
3. Paterson DR. A clinical type of dysphagia. J Laryngol Otol 1919;34:289-291. [CrossRef]
4. Vittal K, Pandian SS, T Malarkodi. Plummer-Vinson syndrome: a case report and medical management. IJSS Case Reports & Reviews 2015;2(6):10-13.
5. Swami H, Ramakrishnan R. Post cricoid web excision. Med J Armed Forces India 2009;65(1):69-70. [CrossRef]
6. Beyler AR, Yurdaydin C, Bahar K, Gören A, Soykan I, Uzunalimoglu O. Dilation therapy of upper esophageal webs in two cases of Plummer-Vinson syndrome. Endoscopy 1996;28(2):266-267. [CrossRef]
7. Yukselen V, Karaoglu AO, Yasa MH. Plummer-Vinson syndrome: a report of three cases. Int J Clin Pract 2003;57(7):646-648.
8. Wynder EL, Hultberg S, Jacobsson F, Bross IJ. Environmental factors in cancer of the upper alimentary tract. A Swedish study with special reference to Plummer-Vinson (Paterson-Kelly) syndrome. Cancer 1957;10(3):470-487. [CrossRef]
9. Chen TS, Chen PS. Rise and fall of the Plummer-Vinson syndrome. J Gastroenterol Hepatol 1994;9(6):654-658. [CrossRef]
10. Basak M, Poyraz T, Ciltas A, Acikgoz O, Osman N, Kutlu M, et al. Plummer-Vinson syndrome. Bakırköy Tıp Dergisi/Medical Journal of Bakirkoy 2006;2(3):109-112.
11. Demirci F, Savas MC, Kepkep N, Okan V, Yilmaz M, Buyukberber M, et al. Plummer-Vinson syndrome and dilation therapy: a report of two cases. Turk J Gastroenterol 2005;16(4):224-227.
12. Chaudhry MA, Alamgir A, Obaid A. Esophagoscopy in the early detection and treatment of Plummer Vinson Syndrome. Journal of Rawalpindi Medical College 2008;12(1):37-40.
13. Salihoun M, Kabbaj N, Raissouni F, Chaoui Z, Mohamed A, Naima A. Safety and Effectiveness of Endoscopic Savary-Gillaard Bougies Dilation in Moroccan Plummer-Vinson Syndrome Patients. ISRN Endoscopy 2013 [Internet] Article ID 137895, 3 pages. [CrossRef]
14. Kantarceken B, Cetinkaya A, Citirik C. Plummer-Vinson sendromu için alışılmadık yaş ve cinsiyet: Olgu bildirimi. Akademik Gastroenteroloji Dergisi 2008;7(2):118-120.
15. Dinler G, Tander B, Kalayci AG, Rizalar R. Plummer-Vinson syndrome in a 15-year-old boy. Turk J Pediatr 2009;51(4):384-386.
16. Samad A, Mohan N, Balaji RVS, Augustine D, Patil SG. Oral manifestations of Plummer-Vinson syndrome: a classic report with literature review. J Int Oral Health 2015;7(3):68-71.
17. Hasan S, Khan NI, Siddiqui A. Plummer Vinson syndrome—a premalignant condition – an overview of literature. Unique Journal of Medical and Dental Sciences 2013;1(1):28-30.
18. Lanke G, Koduru P, Bhutani MS. Plummer-Vinson syndrome presenting as squamous cell carcinoma of esophagus. Journal of Digestive Endoscopy 2016;7(2):71-73. [CrossRef]
19. Larsson LG, Sandström A, Westling P. Relationship of Plummer-Vinson disease to cancer of the upper alimentary tract in Sweden. Cancer Res 1975;35(11 pt. 2):3308-3316.