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Plummer-Vinson syndrome and dilation therapy: A report of two cases

Plummer-Vinson sendromu ve dilatasyon tedavisi: İki olgu raporu

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Plummer-Vinson syndrome is known as the association of post-cricoid dysphagia, upper esophageal web, and iron deficiency anemia. Although correction of iron deficiency may result in resolution of dysphagia and sometimes disappearance of the webs, dilation therapy is usually necessary to remove webs and relieve dysphagia. We report two cases of Plummer-Vinson syndrome. Both patients presented with significant and long-standing dysphagia, sideropenia, glossitis and koilonychia. Our two patients had occasional choking and aspiration episodes at eating and endoscope did not pass through at the level of the upper esophagus. Patients' esophagograms revealed the presence of webs in part of the post-cricoid region. Both patients were treated with esophageal bougienage or balloon dilation, and iron supplementation. The patients were examined periodically for two years after the initial treatment and found to be in good general condition.

Key words: Plummer-Vinson syndrome, dysphagia, upper esophageal web, dilation therapy

Plummer-Vinson sendromu post-krikoid disfaji, üst özofagus vebi ve demir eksikliği anemisinin birlikteliği olarak bilinir. Her ne kadar demir eksikliğinin düzeltilmesi, disfajinin rahatlamasıyla ve bazen de vebin ortadan kalkmasıyla sonuçlansa da genellikle, vebi yok etmek ve disfajiyi rahatlatmak için dilatasyon tedavisi gereklidir. Biz iki Plummer-Vinson sendromu vakası bildirdik. Her iki hasta, önemli ve uzun süren disfaji, sideropeni, glossit ve kaşık tırnak şikayetleri ile müracaat etti. İki hastamızda da bazen yemek yerken boğulma hissi ve aspirasyon periyodları vardı ve endoskop üst özofagustan ileri gitmedi. Hastaların özofagogramları post-krikoid bölgede vebi gösteriyordu. Her iki hasta özofagal buji yada balon ile birlikte demirle tedavi edildi. Tedaviden sonra iki yıl boyunca düzenli muayaneler yapıldı ve hastalar iyi durumda bulundu.

Anahtar kelimeler: Plummer-Vinson sendromu, disfaji, üst özofagus vebi, dilatasyon tedavisi

INTRODUCTION

Plummer-Vinson syndrome is characterized by cervical dysphagia, iron deficiency anemia, and upper esophageal web or webs. While this syndrome is known as Plummer-Vinson syndrome in the United States, it is known as Paterson-Brown Kelly syndrome in the United Kingdom. It has been known since the beginning of the 20th century. Plummer (1) established the syndrome for the first time in 1912 and has published 21 cases with diffuse dilation of the esophagus and spasm of the upper esophagus without anatomic stenosis. Then Vinson (2), Plummer's pupil, published another case with angulation of the esophagus. Paterson and Kelly (3, 4) described for the first time the characteristic clinical signs of the syndrome: anemia, dysphagia, glossitis, cheilitis, iron deficiency, and koilonychia.

Herein we present two patients with significant and long-standing dysphagia and sideropenia. Both patients were treated with dilation therapy and iron supplementation.

CASE REPORTS

Case 1

A 47–year-old woman presented with intermittent and long-standing (five years) symptom of dysphagia. She had a history of five births and two abor-

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tions. The patient informed us that she sometimes took iron salts because of anemia of iron deficiency. The clinical examination showed signs of iron deficiency such as cheilitis and spoon-shaped nails.

Hematological tests showed the presence of an iron deficiency anemia (serum Fe 40 µg/dl, ferritin 50 ng/ml, Hb 9.6 g/dl, Ht 29%). The examination of peripheral blood smear revealed hypochromia and aniso-poikilocytosis. Tests for other causes of chronic anemia were negative. The patient's esophagogram revealed the presence of a web in the subcricoid region (Figure 1) and the endoscope did not pass through at the level of the web. The patient was treated with oral ferrous agent, 150 mg daily (p.o.) for three weeks, in addition to esophageal dilation with Eder-Pustov dilators. After dilation, the endoscope passed through the esophagus easily, but there were remnants of the web (Figure 2). The remaining esophagus, stomach and duodenum were normal. The patient was given iron salt for four months until the hematological tests normalized. The remnant webs disappeared in the follow-up upper endoscopic examination. Two years later the patient was found to be well and she continues to be healthy.

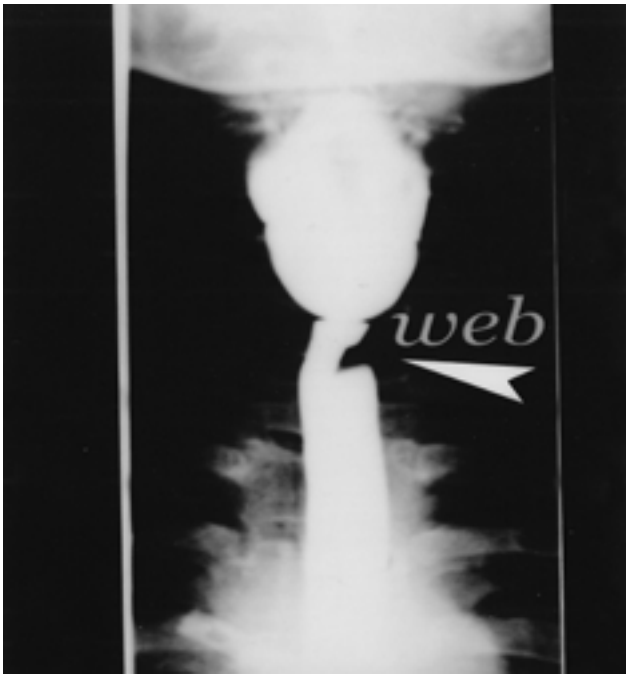

Figure 1. Photographic image of a web (case 1)

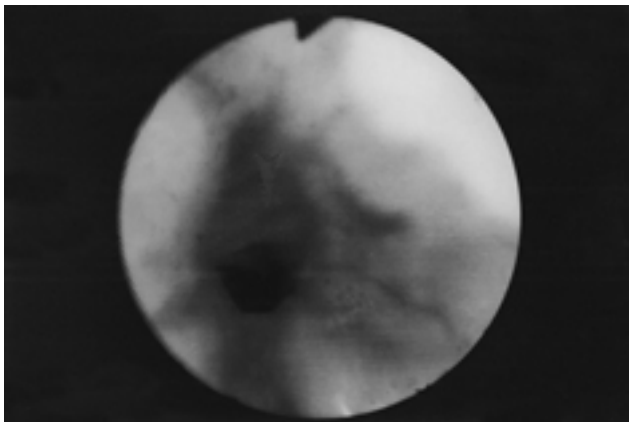

Figure 2. Endoscopic image of web remnants after bougienage dilation (arrow, case 1)

Case 2

A 45-year-old woman presented with progressive dysphagia and repeated episodes of menorrhagia over the preceding three-year period. The patient reported that she sometimes experienced choking and aspiration at eating. The clinical examination revealed signs of iron deficiency anemia, such as glossitis and spoon-shaped nails. Ultrasonographic examination of the lower abdomen showed normal uterus and the diagnostic curettage was also normal.

Esophagogram showed a ring in part of the upper esophagus. The esophagoscopy confirmed the presence of a web in the sub-cricoid region and the instrument did not pass through at the level of the web (Figure 3).

Hematological tests showed signs of iron deficiency anemia including hypochromia, microcytosis (serum Fe: 47 g/dl, serum ferritin: 52 ng/ml, Ht 33%, Hb 10.8 g/dl, MCHC 28%). The examination of peripheral blood smear revealed the presence of hypochromia, poikilocytosis and target cells. The patient was investigated for other causes of chronic anemia, and all results were negative.

The patient was treated with oral ferrous agent, 150 mg daily (p.o.) for three weeks, in addition to esophageal dilation with a balloon. Balloon dilation was successful under scopic control. After balloon dilation, endoscope passed through the esophagus easily (Figure 4) and the remainder of the esophagus, stomach and duodenum were normal.

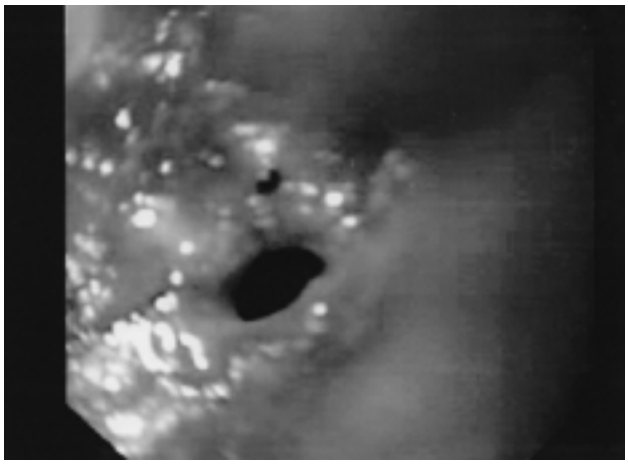

Figure 3. Endoscopic image of esophageal web (case 2)

She continued the intake of iron until her hematocrit and ferritin levels were normalized, which occurred after six months. The patient was found to be in good general condition two years after diagnosis.

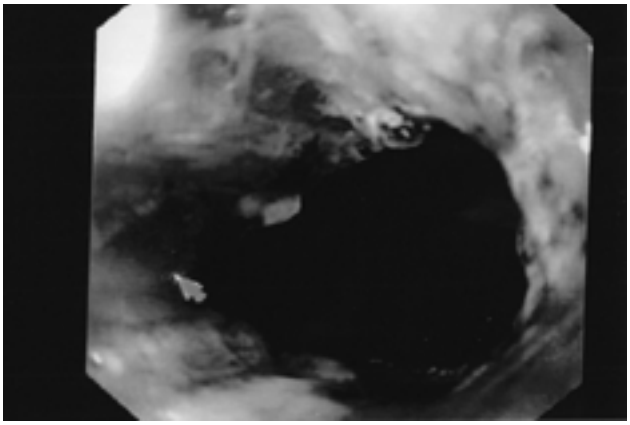

Figure 4. Appearance of upper esophagus after balloon dilation (case 2)

DISCUSSION

The classical triad of the Plummer-Vinson or Paterson-Kelly syndrome consists of upper esophageal web or webs, dysphagia and sideropenic anemia. In the past, this syndrome was more common among the Scandinavian population, especially in rural areas of Sweden, but it is extremely rare today. The decline in reports and incidence of the syndrome may be related to the improvement in nutritional status and better treatment of iron deficiency (5). In Turkey, however, many Plummer-Vinson syndrome cases are still being reported (6-8).

Cervical dysphagia is the rarest type of dysphagia. The most common benign cause of cervical type of dysphagia is upper esophageal web(s). These webs may be found in approximately 5-15% of patients with dysphagia. Webs are fragile membranes and respond well to esophageal dilation therapy (9).

In our cases, dysphagia was the main symptom that led both patients to seek medical help and dilation therapy.

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 (10, 11). Prevalence data are not reliable because many studies done on patients with dysphagia have not included the hematological parameters (12).

The pathogenesis of the syndrome remains unclear, but 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. As a result, mucosal atrophy leads to development of webs (9). It was reported that in a patient with Plummer-Vinson syndrome, iron deficiency caused esophageal motility decrease; new motility studies showed normal amplitude of contraction after iron therapy (13). In another patient with Plummer-Vinson syndrome, iron therapy alone resolved the patient's symptom, and the patient's esophagogram showed 30% benign upper esophagal stricture, reduced from 90% before therapy (14). But population studies do not confirm the etiologic association between iron deficiency and web formation. Furthermore, this theory does not explain the presence of these webs exclusively in the upper part of the esophagus (9, 15). Finally, the precise role of iron deficiency in Plummer-Vinson syndrome has yet to be defined and remains a subject of debate.

Genetic transmission has gained little acceptance to date. But some families and relatives have been reported as having this syndrome. Furthermore, Plummer-Vinson syndrome and sideropenic dysphagia are rare among blacks (9, 16).

Plummer-Vinson syndrome may be accompanied by pernicious anemia, thyroiditis or celiac disease.

Celiac disease is a recognized cause of chronic iron deficiency and should be considered as an etiological factor in sideropenic dysphagia (17-20).

The most important symptom of this syndrome is dysphagia, which is limited to solid foods and is generally intermittent. Patients may also complain about choking and aspiration episodes (21). The symptoms of sideropenic anemia such as weakness, pallor, fatigue and tachycardia may dominate the clinical picture. The clinical signs of sideropenic anemia including angular cheilitis, glossitis and koilonychia may also be seen on physical examination.

The treatment of Plummer-Vinson syndrome is iron supplementation, and iron therapy may be necessary even though hematologic parameters are normal in the presence of a web formation (13, 14). The patients who have choking and aspiration episodes need dilation therapy together with iron supplementation. Endoscopic dilation is simple and a chosen procedure in treatment of the syndrome and cervical web of the esophagus (6, 7). Our two patients had choking and aspiration episodes and the endoscope did not pass through at the level of the web.

It is important that this syndrome be differentiated 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 and surveillance endoscopy is recommended (22, 23). Celiac disease, large diaphragmatic hernia, gastric cancer, Sjogren's syndrome, and pernicious anemia may cause Plummer-Vinson syndrome. It is proposed that these diseases may cause iron deficiency and then result in upper esophageal webs (17-20, 24, 25).

In our patients, treatments were achieved by dilation and iron therapy. Although bougienage dilation was preferred in previous reports (6, 7), we experienced that balloon dilation was efficient and reliable.

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