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A Case of Plummer-Vinson Syndrome Showing Rapid Improvement of Dysphagia and Esophageal Web after Two Weeks of Iron Therapy

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Key Words

Plummer-Vinson syndrome · Iron therapy · Dysphagia · Esophageal web · Iron deficiency anemia

Abstract

Plummer-Vinson syndrome (PVS) is a rare entity characterized by upper esophageal webs and iron deficiency anemia. We report a case of PVS whose esophageal web was rapidly improved by iron therapy. A 77-year-old woman was admitted to our hospital with complaints of dysphagia, vomiting, shortness of breath and weight loss for 1 month. Physical examination revealed conjunctival pallor, koilonychia, angular cheilitis and smooth tongue, and laboratory findings were consistent with microcytic hypochromic anemia with iron deficiency. Gastrointestinal endoscopy and barium-swallow esophagography detected a web that prevented passage of the endoscope into the upper portion of the esophagus. The patient received oral iron therapy daily; the hemoglobin concentration rose to 8.9 g/dl and the complaints of dysphagia were dramatically improved after 2 weeks, with improvement of luminal stenosis confirmed by gastrointestinal endoscopy and barium-swallow esophagography. The PVS described in this report had a distinct clinical course, showing very rapid improvement of dysphagia and esophageal web after 2 weeks of oral iron therapy.

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Introduction

Plummer-Vinson syndrome (PVS), also called Paterson-Brown-Kelly syndrome, is a rare entity whose main clinical features are upper esophageal web(s), dysphagia and iron deficiency anemia [1]. PVS is relatively common among middle-aged female Caucasians in northern countries whereas in Japan, PVS seems to be rare. The report by Uchida et al. [2] demonstrated that PVS was found in 6 (1.7%) of 353 Japanese patients with iron deficiency anemia.

Although the pathogenesis of PVS remains largely unknown, the most probable mechanism of PVS is iron deficiency [3], leading to rapid loss of iron-dependent enzymes due to its high cell turnover. Loss of these enzymes causes mucosal degenerations, atrophic changes and web formation, which are associated with dysphagia. It has been suggested that dysphagia associated with PVS is usually preceded by iron deficiency and is gradually relieved by iron supplements [4]. However, there are some conflicting reports on esophageal webs and latent iron deficiency suggesting no correlation between the same [5]. Indeed, there have been several cases in whom the dysphagia did not respond to iron therapy and ultimately required endoscopic dilatation or incision [6–8].

In this case report, we present a rare case of PVS who showed very rapid improvement of esophageal web and dysphagia after 2 weeks of oral iron therapy.

Case Report

A 77-year-old woman was admitted to Fujita Health University Hospital with complaints of dysphagia and vomiting after every meal, shortness of breath and 5 kg of weight loss for 1 month. Physical examination revealed conjunctival pallor, koilonychia, angular cheilitis and smooth tongue with loss of the normal tongue papillae (fig. 1).

Laboratory findings were consistent with microcytic hypochromic anemia with iron deficiency: red blood cell count $274 \times 10^4/\mu\text{l}$, hemoglobin 3.8 g/dl, hematocrit 28.1%, mean corpuscular volume 57.0 fl, mean cell hemoglobin 13.9 pg, mean cell hemoglobin concentration 24.4%, serum iron level 7 $\mu\text{g/dl}$, total iron binding capacity 425 $\mu\text{g/dl}$, and serum ferritin level 2.0 ng/ml. Serum liver and kidney function tests were unremarkable (serum AST 13 IU/l, serum ALT 5 IU/l, serum BUN 14.7 mg/dl and serum creatinine 0.49 mg/dl).

After erythrocyte suspension, gastrointestinal endoscopy was attempted. A web was detected that prevented passage of the endoscope into the upper portion of the esophagus (fig. 2a). Then we used transnasal endoscopy and passage of transnasal endoscopy through the web was successful. Barium-swallow esophagography also revealed a circumferential web at the cervical esophagus (fig. 3a). Computed tomography revealed no abnormality around the esophagus, such as a tumor or swollen lymph nodes, which can cause luminal stenosis. No clear cause of the anemia was identified. Because of the upper esophageal webs, iron deficiency anemia, angular cheilitis and smooth tongue with loss of normal tongue papillae, PVS was diagnosed. The patient received oral iron therapy daily; the hemoglobin concentration rose to 8.9 g/dl and the complaints of dysphagia were dramatically improved after 2 weeks. Gastrointestinal endoscopy and barium-swallow esophagography confirmed the improvement of luminal stenosis (fig. 2b, fig. 3b).

Discussion

The pathogenesis of PVS remains largely unknown. However, the most probable mechanism of PVS is iron deficiency [3]. The other causes, genetic, environmental and immunological factors have not been proven to play an exact role in the pathogenesis of this syndrome, although celiac disease, thyroid disease and rheumatoid arthritis have been reported with PVS [1, 9, 10]. Regardless of its source, the theory is based on iron deficiency [3], which leads to rapid loss of iron-dependent enzymes. Loss of these enzymes cause web formation and eventually lead to cancer development of the upper gastrointestinal tract [11, 12]. Tissue iron plays an important role in the proliferation of epithelial cells. The physical signs of tissue iron deficiency include smooth tongue, angular cheilitis and koilonychia, which were also observed in our patient. Moreover, the epithelial layer of the upper alimentary tract is especially susceptible to iron deficiency because of its high cell turnover.

It has been suggested that dysphagia associated with PVS is improved by iron supplements [4], while there have also been several cases in whom the dysphagia did not respond to iron therapy and ultimately required endoscopic dilatation or incision [6–8]. Notably, our case showed very rapid improvement of dysphagia after 2 weeks of oral iron therapy. Gastrointestinal endoscopy and barium-swallow esophagography also confirmed the improvement of luminal stenosis within the same period. Since the correlation between esophageal webs and latent iron deficiency has not been conclusive [5], the features of cases that will be resolved by iron therapy alone need to be clarified. Moreover, esophageal webs may relapse if iron deficiency recurs; therefore, careful follow-up is mandatory for these patients. Finally, PVS is associated with an increased risk of upper alimentary tract cancers [11, 12]. Endoscopic surveillance is also necessary because of the risk of cancer.

Disclosure Statement

No conflicts of interest exist.

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Fig. 1. Angular cheilitis and smooth tongue with loss of the normal tongue papillae.

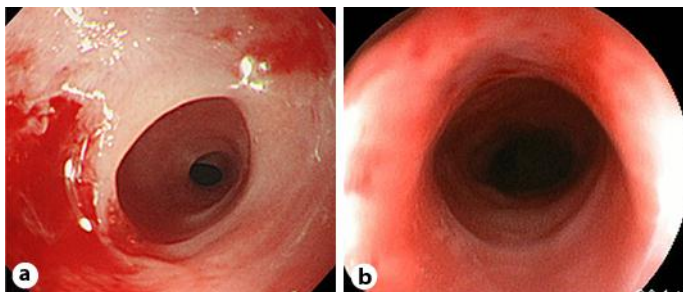


Fig. 2. Upper esophageal web seen on gastrointestinal endoscopy before (a) and after 2 weeks of iron therapy (b).

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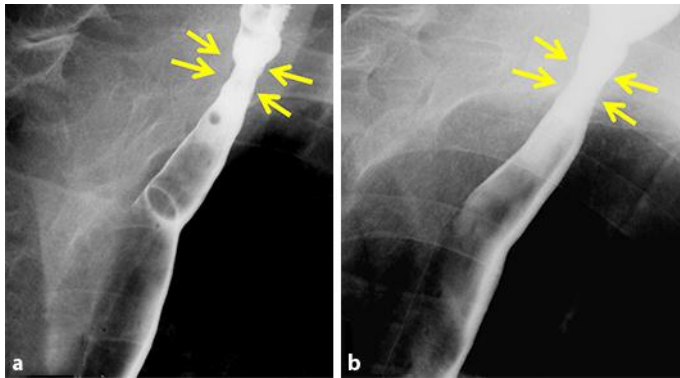


Fig. 3. Upper esophageal web seen on barium-swallow esophagography indicated by arrows before (a) and after 2 weeks of iron therapy (b).