



**INTERNATIONAL JOURNAL OF  
PHARMACEUTICAL SCIENCES**  
[ISSN: 0975-4725; CODEN(USA):IJPS00]  
Journal Homepage: <https://www.ijpsjournal.com>



## Case Study

# Plummer-Vinson Syndrome; A Rare Case Report

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## ARTICLE INFO

Received: 15 May 2024

Accepted: 19 May 2024

Published: 04 June 2024

### Keywords:

Plummer, Vinson, Syndrome,  
Rare

### DOI:

10.5281/zenodo.11482508

## ABSTRACT

Plummer-Vinson syndrome (PVS) is a rare disorder that causes dysphagia, glossitis, and esophageal webs. It has been associated with chronic severe iron deficiency anemia, although the fact that its pathophysiology is still completely unknown. We report a case of 45-year-old woman was admitted to the Hospital with complaints of throat pain, shortness of breath, dysphagia and Physical examination revealed koilonychia, angular cheilitis, and smooth tongue. laboratory findings were consistent with microcytic hypochromic anemia with iron deficiency, Gastrointestinal endoscopy. The patient received iron therapy and the hemoglobin concentration rose to 8.5 g/dl and the complaints of dysphagia were dramatically improved after 2 weeks

## INTRODUCTION

Plummer-Vinson syndrome (PVS) is a rare disorder that causes dysphagia, glossitis, and esophageal webs. It has been associated with chronic severe iron deficiency anemia [1]. It has the names of two physicians from America, Porter Paisley Vinson and Henry Stanley Plummer. Named for two British otolaryngologists, Drs. Adam Brown-Kelly and Donald Ross Paterson, PVS is also referred to as Kelly-Paterson syndrome [2]. Iron deficiency is the most common cause of PVS, although the fact that its pathophysiology is still completely unknown [3],

Because of its high cell turnover, it causes a rapid depletion of iron-dependent enzymes. Mucosal degenerations, atrophic alterations, and web formation brought on by the loss of these enzymes have been associated to dysphagia. According to certain theories, iron deficiency frequently precedes dysphagia related to PVS, and iron supplements progressively alleviate the condition [4]. However, conflicting assessments about esophageal webs and latent iron deficiency indicate that there is no discernible association between the two. It is accurate that in several patients, endoscopic dilatation or incision was

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Relevant conflicts of interest/financial disclosures: The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.



necessary since the dysphagia did not improve with iron therapy [5]. This case report presents a rare case of PVS where, following two weeks of oral iron therapy, the patient's dysphagia and esophageal web improved very quickly.

### CASE PRESENTATION

A 45-year-old woman was admitted to the Hospital with complaints of throat pain since 1 year, shortness of breath, she mentioned having dysphagia with only solid foods since 1 year. On physical examination, her vitals are normal. She has limited mouth opening, koilonychia, angular cheilitis, and smooth tongue, pigments on both cheeks and nasal bridge, perioral.

Laboratory findings were consistent with microcytic hypochromic anemia with iron deficiency: red blood cell count  $3.63 \times 10^6/\mu\text{l}$ , hemoglobin 5.9 g/dl, hematocrit 22.4%, mean corpuscular volume 57.0 fl, mean cell hemoglobin 13.5pg, mean cell hemoglobin concentration 26.5%, serum iron level 7  $\mu\text{g/dl}$ , serum ferritin level 4.5 ng/ml, vitamin B12 480 pg/ml. A web was found to be obstructing the endoscope's ability to enter the upper part of the esophagus. Then we utilized trans nasal endoscopy, and it was successfully passed through the web. An additional finding from barium-swallow esophagography was a circumferential web at the cervical esophagus. No abnormalities surrounding the esophagus, such as a tumor or enlarged lymph nodes that could induce luminal stenosis, was detected by computed tomography. Because of the iron deficiency anemia, upper esophageal webs, dysphagia and physical examination, Plummer-Vinson Syndrome was diagnosed. She was transfused with 1 unit of packed red blood cells and later given parenteral dextran for her iron deficiency. She was prescribed with the medication iron sucrose in 100ml NS intravenously once alternative days, vitamin C 500mg orally twice a day, multivitamin orally once a day. the hemoglobin concentration rose to

8.5 g/dl and the complaints of dysphagia were dramatically improved after 2 weeks. Rupture and mechanical dilation of the web with an endoscope can be carried out, as it was with our patient, in the event of substantial esophageal obstruction caused by several esophageal webs or chronic dysphagia despite medicinal treatment.

### DISCUSSION

PVS was suspected in this patient who had dysphagia and iron deficiency anemia; further barium swallowing confirmed the diagnosis. The syndrome is a rare condition marked by dysphagia, esophageal webs, and iron-deficiency anemia.[6] Most of the pathophysiology of PVS is still unknown. However, Iron deficiency anemia is the most common mechanism of PVS. Although rheumatoid arthritis, thyroid disorders, and celiac disease have been linked to PVS, the precise roles played by the other causes—genetic, environmental, and immunological factors—in the etiology of this syndrome remain unknown. [7] This causes iron-dependent enzymes to rapidly disappear. Loss of these enzymes results in the formation of webs, which ultimately cause upper gastrointestinal tract cancer. The proliferation of epithelial cells is significantly aided by tissue iron. The physical manifestations of angular cheilitis, koilonychia, and smooth tongue are indicative of tissue iron insufficiency, and our patient exhibited all three [8]. In addition, because of its fast cell turnover, the epithelial layer of the upper gastrointestinal tract is particularly vulnerable to iron deficiency. Iron-dependent oxidative enzymes cannot perform at their best in patients with iron shortage, and dependent metabolic pathways including oxidative phosphorylation are affected. By encouraging anaerobic metabolism, this causes myasthenic alterations in the esophageal muscles, which in turn creates esophageal webs. Esophageal muscle dyscoordination is rarely the cause of dysphagia [9]. It has been suggested that dysphagia



associated with PVS is improved by iron supplements [4]. However, there have also been several instances when the dysphagia did not improve with iron therapy, necessitating an endoscopic dilation or incision [10]. Notably, after two weeks of oral iron therapy, our patient demonstrated a quick improvement in dysphagia. The improvement of luminal stenosis within the same period was also confirmed by barium-swallow esophagography and gastrointestinal endoscopy. Given that there is still no solid evidence linking latent iron deficit to esophageal webs [11]. It's important to define the characteristics of cases that iron therapy alone will be able to treat. Furthermore, patients with iron deficiency must have close monitoring because recurrence of esophageal webs is possible. Lastly, a higher incidence of upper gastrointestinal tract malignancies is linked to PVS. The potential risk of malignancy necessitates endoscopic surveillance in addition [12].

## CONCLUSION

PVS is a rare triad that develops iron-deficiency anemia, esophageal webs, and increasing dysphagia in the same patient. Early replacement of iron stores and management of the esophageal webs of PVS are essential because the webs can develop into pharyngeal or esophageal squamous cell carcinoma. In the case of our patient, her dysphagia evolved while on iron supplementation.

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**HOW TO CITE:** Shaik Khadeer Ahamad, Deepthi Dara, Sanjana Reddy Thota, Rama Rao Tadikonda, Plummer-Vinson Syndrome; A Rare Case Report, Int. J. of Pharm. Sci., 2024, Vol 2, Issue 6, 230-233. <https://doi.org/10.5281/zenodo.11482508>

