Plummer–Vinson syndrome: A rare case of dysphagia in a middle aged female

Nicholas Figaro, Keegan Figaro, Sanjeev Solomon, Solaiman Juman

CASE REPORT

A 42-year-old female presented to the Otolaryngology Department with a two year history of progressive dysphagia to solids. She did not have any comorbidities yet complained of fatigue and shortness of breath on exertion. When questioned about her menstrual cycle, the patient was unaware that she was having menorrhagia for several years. Her physical examination was normal with the exception of her skin pallor and platynychia (Figure 1). Hematological investigation revealed a hemoglobin level of 6.5 g/dL, hematocrit of 24%, mean corpuscular volume of 58.5 fL, a red cell distribution width of 19.1%, and a serum ferritin level of 9 ng/mL. Barium esophagogram demonstrated an esophageal web within the proximal esophagus at the level of the C6 vertebral body (Figure 2). A diagnosis of Plummer–Vinson syndrome was made and the patient was referred to gastroenterologist for therapeutic intervention.

Upper gastrointestinal endoscopy showed a focal, tight upper esophageal web (Figure 3). Serial dilation was performed using 11, 13, and 15 mm Savary-Gilliard dilators over a guidewire. Adequate dilation was successfully achieved despite moderate resistance with the last two dilators. Post-dilation inspection showed a superficial tear with no signs of bleeding or perforation (Figure 4). The patient had an uneventful post-procedural course and her dysphagia was relieved immediately. She was discharged with oral hematinics.

The patient was evaluated six weeks post-procedure at the outpatient clinic. She had no swallowing complaints and histopathological analysis of the duodenal biopsy specimens revealed no evidence of celiac disease. She was counseled on the risk of post-cricoid carcinoma and the need for long-term surveillance of the upper gastrointestinal tract. Gynecological consultation was requested to investigate the patient’s menorrhagia. A bulky fibroid uterus was discovered during the assessment, for which the patient eventually had a total hysterectomy.
DISCUSSION

Plummer–Vinson syndrome (PVS) or sideropenic dysphagia is an uncommon entity characterized by dysphagia, iron deficiency anemia, and the presence of an esophageal web [1]. The frequency of reported cases has decreased which may possibly be due to the overall improvement in nutrition and health care [2]. The syndrome occurs more commonly in middle-aged women, approximately 9 times more frequently than men [2, 3]. Although the exact etiopathogenesis of this entity is unclear, several hypotheses have been made, the most popular of which is based on iron-dependent oxidative enzyme deficiency [2, 4]. These iron-dependent oxidative enzymes are crucial for maintaining the epithelial integrity of the upper alimentary canal. As a result, deficiencies of these enzymes result in gradual degradation of pharyngeal musculature, leading to mucosal atrophy and web formation [4, 5]. However, studies have shown that PVS is not naturally endemic in areas of iron deficiency anemia, in fact, only 10% of patients with iron deficiency anemia develop PVS. This suggests that other factors like genetic, environmental, and autoimmune may be involved [2, 6].

Holistic management of the patient with PVS includes investigating and treating the etiology of the chronic iron deficiency anemia, correcting the anemia, and resolving the symptom of dysphagia. Common causes of iron deficiency anemia include gastrointestinal bleeding, nutritional deficiency due to poor intake or malabsorption, gastrointestinal parasites, menorrhagia, and menometrorrhagia [7].

Iron supplementation and mechanical dilation are the mainstay of treating the dysphagia of PVS [8]. Correction of anemia via oral or intravenous route may be sufficient to eradicate the web and resolve dysphagia in patients who are not considerably obstructed. Patients who have failed conservative hematinic management and others who are inherently obstructed like the index case will require endoscopic therapeutic intervention [3]. Plummer–Vinson syndrome is considered a premalignant condition and as such, long-term follow-up of the post-cricoid region is mandatory [1, 3, 9].

CONCLUSION

Plummer–Vinson syndrome is a rare condition that is defined by the triad of dysphagia, chronic iron deficiency anemia, and the presence of an esophageal web. Management of PVS includes identifying and correcting the underlying cause of the iron deficiency anemia, treating the anemia with hematinics and mechanically resolving the dysphagia as required. Long-term follow-up of these patients are necessary as PVS is considered a premalignant condition for gastrointestinal malignancies.

Keywords: Dysphagia, Esophageal web, Iron deficiency anemia, Plummer–Vinson syndrome

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REFERENCES


Author Contributions
Nicholas Figaro – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved
Keegan Figaro – Acquisition of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved
Sanjeev Solomon – Acquisition of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved
Solaiman Juman – Analysis of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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