

Case Report

Dysphagia in a middle - aged female : A Case report.

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Abstract

Plummer-Vinson syndrome (PVS) is characterized by a hypopharyngeal or postcricoid web causing progressive dysphagia and iron deficiency anemia. We report the case of a middle-aged female who presented to us with complaints of easy fatigability and progressive dysphagia mainly to solids for six months. The patient had marked pallor. Her upper endoscopy showed hypopharyngeal web, which was confirmed by barium swallow study. A diagnosis of Plummer-Vinson syndrome was made. The patient was initiated on oral iron supplementation with marked improvement in her overall well being and a steady improvement in her dysphagia.

Key words: Dysphagia, Iron deficiency anaemia, Hypopharyngeal web, Plummer-Vinson syndrome.

Introduction

Plummer Vinson syndrome or Paterson-Brown Kelly syndrome consists of a triad of upper esophageal web, dysphagia, and iron deficiency anemia. Majority of the affected patient are Caucasian middle-aged females. Plummer-Vinson syndrome is a precancerous condition and it is estimated that 3% to 15% of the affected individuals are at risk of developing esophageal or pharyngeal carcinoma.

Case History

A 34-year-old female presented with chief complaints of progressive dysphagia, mainly to solids for the past six months with easy fatigability. There was no history of loss of appetite or loss of weight. No history of fever, nausea, or vomiting. No history of altered bowel habits, melena, or hematochezia. No history of ingestion of NSAIDs or other drugs. There was no past history of alkaline or acid ingestion.

On Physical examination was significant for marked pallor and angular cheilitis. Her nails were spoon shaped (koilonychia) and brittle. Auscultation of heart revealed hemic murmurs. Per abdomen exam revealed it to be soft and nontender with no organomegaly. Rest of her examination was benign. Her laboratory data showed a hemoglobin of 6.3 gm% with a reticulocyte count of 0.1%. Peripheral smear showed marked microcytic, hypochromic red cells with anisopoikilocytosis.

The patient was subjected to upper endoscopy which showed a postcricoid web and the scope could not be negotiated beyond the web. Her barium swallow under fluoroscopic control showed a linear defect in the anterior wall of the hypopharynx suggestive of a postcricoid web. The patient was started on oral iron supplementations and has been on regular followup ever since. Patient has reported marked improvement in her dysphagia and her hemoglobin at six-month follow up has increased to 9.7.

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Discussion

Plummer-Vinson syndrome (also known as Paterson-Brown Kelly syndrome and sideropenic dysphagia) is named after Henry Stanley Plummer and Porter Poisley Vinson, the staff physicians at Mayo Clinic, who gave a description of this syndrome in 1912. Later in 1919, it was Paterson who drew attention to its association with post-cricoid carcinoma⁽¹⁾. Majority of the reported case have been in white Caucasian females in their fourth to seventh decade who report painless and progressive dysphagia to solids^(1, 2). The characteristic triad of the syndrome consists of dysphagia, postcricoid or upper esophageal webs, and iron deficiency anemia. The other features include angular cheilitis, atrophic glossitis, and koilonychia and brittle nails. Around 30% of the patient may have splenomegaly⁽³⁾.

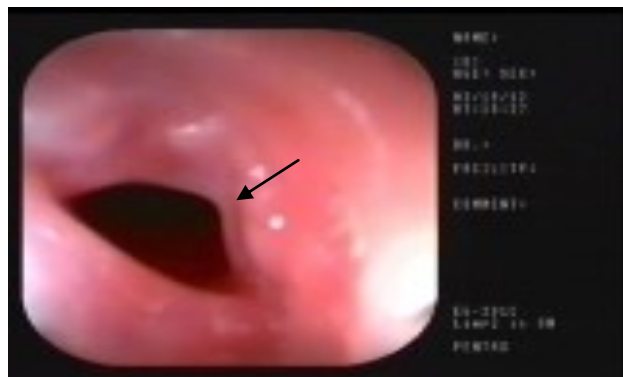


Fig:1 – Esophagogastroduodenoscopy showing the post cricoid web causing narrowing of the esophageal lumen

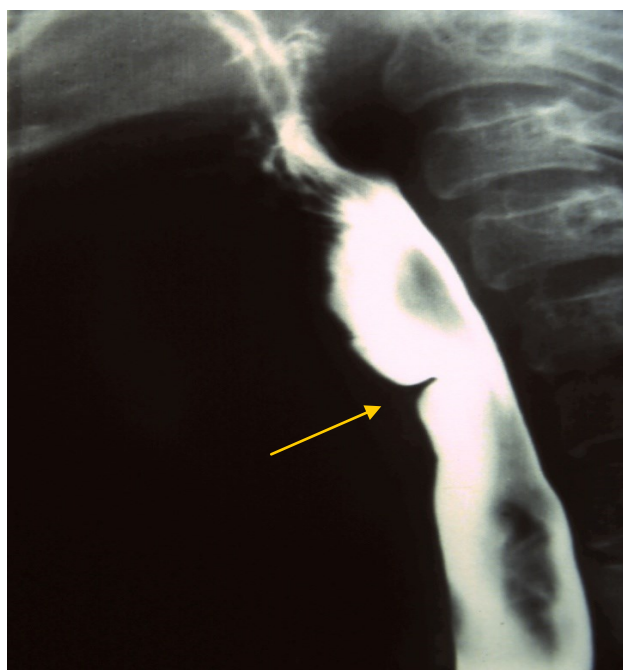


Fig:2 – Barium study showing the esophageal web

Plummer-Vinson syndrome increases the risk of upper alimentary tract cancers, and the incidence of esophageal cancers has been reported to be around 3% to 15%⁽⁴⁾. The exact pathogenesis of this syndrome still eludes the clinicians though iron deficiency, autoimmune etiology, malnutrition, and genetic causes have been proposed. It has been postulated that iron deficiency causes a decrease in the iron-dependent oxidative enzymes resulting in degradation of pharyngeal muscles and causes mucosal atrophy which ultimately results in the formation of webs. This is supported by the fact that these patients show a dramatic improvement when they are supplemented with iron formulations⁽⁵⁾.

Diagnosis is based on the demonstration of esophageal web either by endoscopy or barium swallow studies. Plummer-Vinson syndrome is managed easily with iron supplements and/or mechanical dilatation of the web(s). Patients should be followed closely due to its association with squamous cell carcinoma of the pharynx and esophagus. Yearly surveillance with upper GI endoscopy has been suggested though the effectiveness of such a recommendation is yet to be confirmed⁽⁶⁾.

Conclusion

Plummer-Vinson syndrome refers to a classical triad of upper esophageal web, dysphagia and anemia and seen mainly in white females, the etiopathogenesis of which is poorly understood. Though rare, it is a precancerous condition of the upper gastrointestinal tract, and the patients need regular surveillance endoscopy. Treatment with oral iron supplements is effective in relieving the dysphagia. Some patients may need endoscopic dilation of the web.

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