



A Case Report- Plummer-Vinson Syndrome: A Rare Syndrome in Male

Authors

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Abstract

Plummer-Vinson Syndrome (PVS) also known as Patterson-Brown-Kelly syndrome is a combined presentation of triad of dysphagia, iron deficiency anemia & esophageal webs, a rare syndrome seen more often in middle aged females. Here we present a rare presentation in the young male patient with long standing dysphagia.

Introduction

The association of postcricoid dysphagia, upper esophageal webs and iron deficiency anemia is known as “Plummer-Vinson Syndrome” or “Paterson –Brown Kelly syndrome”. The term “sideropenic dysphagia” has also been used, because the syndrome can occur with iron deficiency (sideropenia), but is not associated with anemia⁽¹⁾. Plummer-Vinson syndrome (PVS) is named after Henry Stanley Plummer and Porter Paisley Vinson, who were physicians on the staff of the Mayo Clinic. In 1912, Plummer reported a series of twenty one patients with long-standing iron deficiency anaemia, dysphagia and spasm of the upper esophagus without anatomic stenosis, which was described as hysterical dysphagia. In 1919 Vinson reported another case of 'angulation' of the oesophagus and attributed the first description of this entity to the earlier report of Plummer⁽²⁾. Another term is Paterson-Brown-Kelly syndrome, named after Donald Ross

Paterson and Adam Brown- Kelly, who were the first to describe the characteristic signs and symptoms (including anemia) of the syndrome independently in 1919. Paterson gave the fullest description but without reference to anaemia. He was also the first to draw attention to an association with post-cricoid carcinoma⁽²⁾. Because of the constant presence of dysphagia associated with the characteristic mucosal lesions and evidence of diminished body iron stores usually associated with low serum iron values, Waldenstrom & Kjellberg introduced the term ‘Sideropenic dysphagia’ to describe this syndrome⁽³⁾.

Case Report

A 25 year old male patient presented to our outpatient department with difficulty of swallowing since 1.5 years, generalized weakness, easy fatigability, shortness of breath since 15 days. Difficulty in swallowing is for morsels of

solid dry food, which made patient shift to soft and semisolid diet since then, and developed loss of appetite.

Physical examination revealed height of 170cm and weight of 74kgs with BMI of 25.60kg/m². Patient had pallor with koilonychia (figure 1). Investigations revealed Haemoglobin of 7.6 gm% with MCV as 65.8fl, MCH as 15.8pg, MCHC 24.1%. Peripheral smear revealed microcytic hypochromic anemia. Serum ferritin levels as 6.20 ng/ml, serum TIBC as 544 microgram/dl, serum vitamin B12 of 1000 pg/ml and folic acid as 5.18 ng/ml. None of the other tests showed abnormalities. Upper gastrointestinal endoscopy and barium swallow was performed to assess the cause of dysphagia. Upper Gastro-intestinal Endoscopy showed constriction at postcricoid region and scope could not be negotiable further (figure 2). On Barium swallow radiograph showed circumferential radiolucent rings arising from the anterior wall in upper esophagus at c5-6 level intervertebral disc with proximal dilatation suggestive of esophageal webs (figure 3, 4).

Fulfilling the classical triad of dysphagia, iron deficiency anemia and postcricoid upper esophageal webs, Plummer Vinson syndrome (or Paterson Brown Kelly syndrome) was diagnosed. Patient was counseled regarding need for regular surveillance for the development of an upper gastrointestinal malignancy and was given 5days of parenteral iron supplementation followed by oral ferrous sulphate. Patient is under regular follow-ups and showed marked improvement with respect to symptoms of dysphagia and fatiguability.



Figure 1: Koilonychia



Figure 2: Upper GI Endoscopy: Postcricoid Esophageal web.

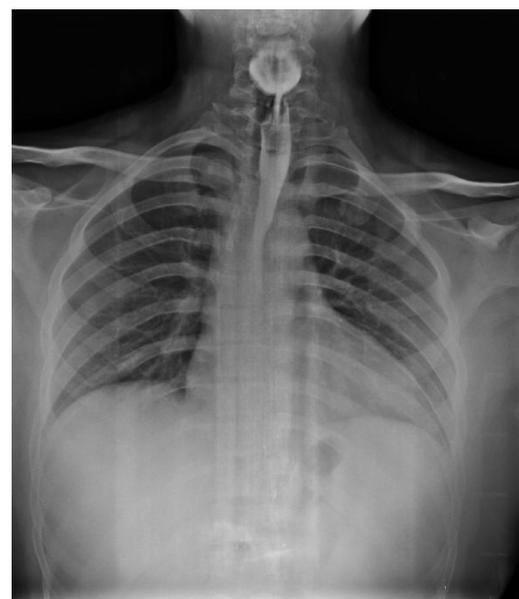


Figure 3: frontal view radiograph: Postcricoid esophageal webs.

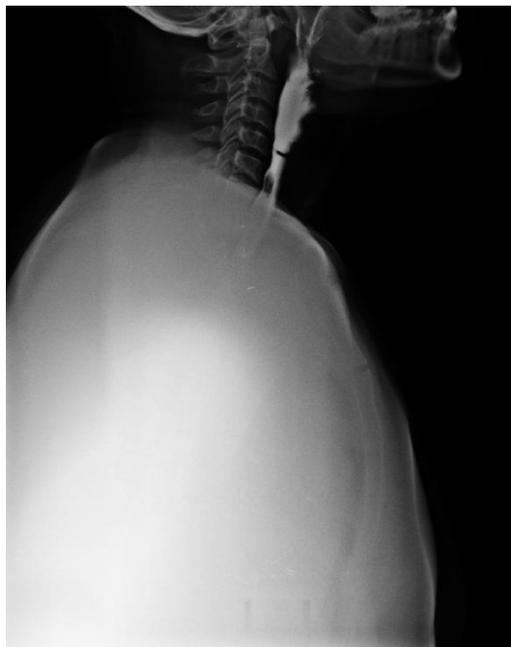


Figure 4: lateral view radiographs: Postcricoid esophageal webs.

Discussion

Plummer Vinson Syndrome (PVS) is a very rare disease. Prevalence has been around less than 1 in 1,000,000. Mostly, middle aged females are affected. The patients usually present with features of anemia like Iron deficiency anemia has been strongly associated with PVS as correction of anemia usually regresses the signs and symptoms of this disorder and also relieves dysphasia in some patients⁽⁴⁾. Genetic predisposition has been linked with PVS as multiple members of the family could be involved. Even autoimmune processes such as celiac disease, rheumatoid arthritis, thyroiditis have been associated⁽¹⁾. Diagnosis is established by confirmation of iron deficiency anemia with demonstration of web by upper GI endoscopy.

The exact etiology is still unclear. The most important possible etiological factor is iron deficiency. On the other hand, the alimentary tract is susceptible to iron deficiency; it rapidly loses iron dependent enzymes due to its high cell turnover, which is speculated to cause mucosa degeneration and web formation⁽⁵⁾. However, large clinical series suggested that for many patients iron deficiency is neither a necessary nor a sufficient cause of web formation⁽⁶⁾. Indeed,

impaired esophageal motility has been described in Plummer-Vinson syndrome and it was corrected by iron treatment. It has been shown that iron deficiency can precede dysphagia. It is thought to be a premalignant condition, and regular follow-up is warranted⁽¹⁾. The symptoms usually resolve by correction of anemia; however, dilatation of the web may be necessary for relieving the dysphagia. Management involves correction of anemia, endoscopic dilatation, balloon dilatation, or incision of oesophageal web. In view of risk for malignant changes in upper gastrointestinal tract, patients require regular follow-up and yearly endoscopic evaluation at least for initial few years after management⁽⁶⁾.

Conclusion

Since Plummer Vinson syndrome is a preventable cause of upper esophageal carcinoma, prediction of dysphagia and questioning regarding dysphagia while taking history in patient presenting with iron deficiency is necessary and can prompt clinician for early intervention. Though literature projects Plummer Vinson syndrome as rare syndrome, seen in women with iron deficiency preceding dysphagia, in the present case patient had dysphagia for 1.5 years and was so trivial that only after iron deficiency anemia symptoms prompted patient for visiting clinician. Hence in patients presenting with iron deficiency, taking history regarding any dysphagia can help clinicians to detect any cases of Plummer Vinson Syndrome and thereby can initiate early interventions & regular surveillance.

References

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