



Plummer Vinson Syndrome in 21- Year-Old Female: A Case Report

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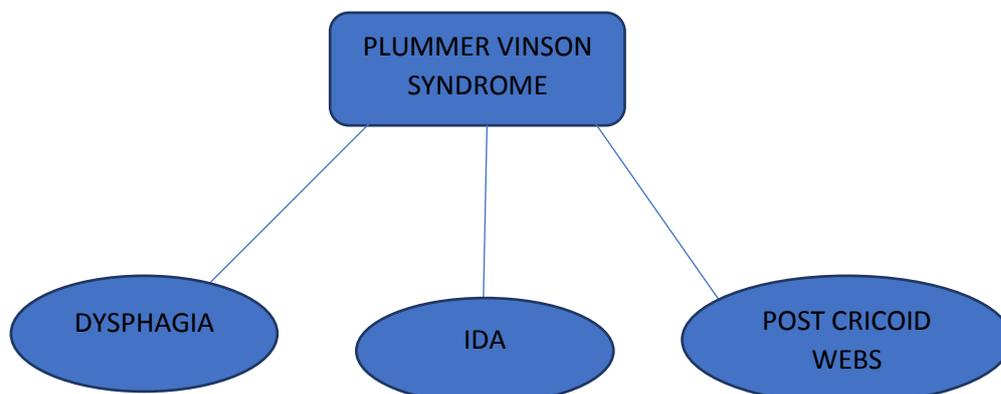
Abstract

PVS is a rare syndrome of dysphagia, iron deficiency anemia, and post cricoid webs. IDA has been hypothesized to play an etiological role. Here we report a case of 21-year-old female who presented to us with shortness of breath, and further evaluation has set forth to an incidental diagnosis of PVS. Many previous studies, elucidated the resolution of dysphagia in most cases with iron therapy alone, but few have shown significant improvement with a combination of web dilatation and iron therapy as observed in our patient.

Introduction

Plummer Vinson Syndrome, also known as Paterson-Brown- Kelly Syndrome is a rare diagnosis, with a combined presentation of Iron deficiency anemia, dysphagia and esophageal webs. It is named after two American Physicians, Dr. Henry Stanley Plummer and Dr. Paisley Vinson. Kelly Patterson syndrome derived its name from two British otolaryngologists, Dr. Adam Brown Kelly and Dr. Donald Roes Paterson¹. Grossly esophageal webs and strictures are pathognomic of PVS. PVS is most frequently reported in middle aged white women and is very

rarely noted in males, children and adolescents.² Lab investigations of PVS are consistent with the findings of Iron deficiency anemia. Barium esophagogram is the best initial study for the diagnosis of PVS, though esophagogastroduodenoscopy is also used to visualize the esophageal webs¹. PVS raises the risk of oral squamous cell carcinoma and esophageal carcinoma at the post cricoidal tissue web³. Waldenstrom and Kjellbug introduced the term sideroblastic dysphagia to describe this syndrome because of the constant association with dysphagia and low iron stores.



Case Report

A 21-year-old female patient, known case of hypothyroidism, presented to EMD with complaints of shortness of breath and generalized weakness since 20 days. After being admitted for further evaluation and management she revealed a history of difficulty of swallowing solid food since childhood. On examination, patient was conscious coherent oriented, and afebrile with vital stables. Locally she had conjunctival pallor with koilonychia. Initial investigations revealed a Hemoglobin level of 3.9gm/dl with MCV-68fm/L, MCH-22picograms, MCHC-.28mg/dl. Serum vitamin b12 and folic acid levels were measured and were 95pg/dl and 10.2ng/dl respectively. Serum ferritin levels were 5.9ng/ml, serum 301microgram/dl. Peripheral smear showed marked anisopoikilocytosis, microcytic hypochromic anemia with tear drop, target cells, and schistocytes. Hematocrit, LFTS, RFTS and PT-INR were within normal limits. She was transfused with 2 units of PRBC in view of low hemoglobin and her post transfusion Hb was 8.8mg/dl. To further workup her source of iron deficiency, her stools were negative for blood and then an Upper gastrointestinal endoscopy was done which revealed post cricoid webs, and endoscope was not negotiable beyond. Upon further gastroenterology consultation esophageal dilatation was performed which resolved her dysphagia.

The patient fulfills the classic triad of Plummer Vinson Syndrome, with Iron deficiency anemia, dysphagia, with post cricoid upper esophageal webs. Patient was conservatively managed with IV INJ FERRINJECT (FERRIC CARBOXYMALTOSATE)/ PARENTAL IRON 1500MG, and other supportive measures. Patient showed drastic improvement clinically and has been under regular follow ups.

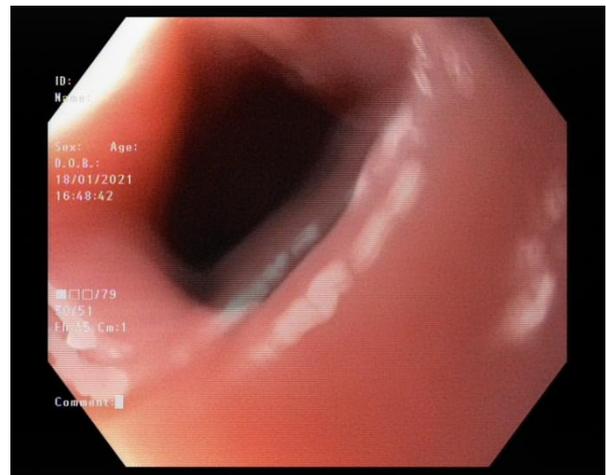


Fig 1: Upper GI Endoscopy

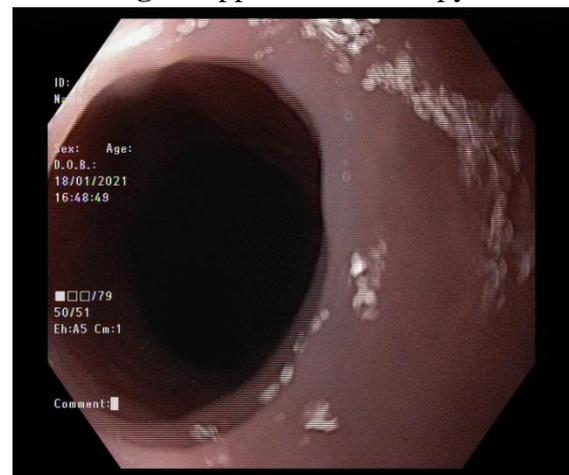


Fig 2: Post Cricoid Esophageal Webs

Discussion

In 1912, Plummer reported a survey of 21 patients with long standing Iron deficiency anemia, dysphagia, and spasm of upper esophagus without anatomic stenosis which was described as hysterical dysphagia. In 1919, Vinson reported another case of angulation of esophagus and attributed the first description of this entity to the earlier report of Plummer. Another term Paterson Kelly Syndrome named after Donald Patterson gave the fullest description without reference to anemia. He was also the first to draw attention to the association with post cricoid carcinoma⁴.

In nearly 20th century, PVS seems to be common in Caucasians of Northern countries, particularly among middle aged perimenopausal women with mean age of presentation being 47 years⁵. Here we report a rare case of 21-year-old female presenting with shortness of breath and fatigue since 20 days. However, some pediatric and adolescent cases

have also been reported⁵. Very few cases of PVS were reported in males as seen in Priya et al study, which reported PVS in a 43-year-old male, and Bentoor et al study, in a 25-year-old male^{4,5}. It is considered that PVS has now become a rare syndrome with an improvement in nutritional status, and availability of Iron supplements.

The exact pathogenesis of Plummer Vinson Syndrome is not clear, but various theories like Iron Deficiency Anemia, malnutrition, genetic predisposition and autoimmune etiologies have been postulated, among which IDA has been the widely accepted, as dysphagia along with webs have shown a significant improvement with iron supplementation. Patients with IDA have rapid loss of iron dependent enzymes due to high cell turnover. The iron dependent oxidative enzymes are unable to function at an optimum level and the dependent metabolic pathways such as oxidative phosphorylation are compromised. This promotes anaerobic metabolism in esophageal muscles forming esophageal webs. A decrease in enzymes leads to esophageal atrophy, decrease in regeneration capacity of the mucosa and stimulates carcinogens and cocarcinogens, which predisposes oral cavity and esophagus to malignancy². A mutation in Tmprss6 gene is shown to lead to PVS. The gene encodes for protein hepcidin, of which when levels increase, the release of iron from ferritin decreases, leading to iron deficiency anemia¹.

Another theory of minimal importance was a deficient level of vit B9 and B12 contributed to PVS. In our case, B9-10.2ng/dl, B12-95pg/dl. Plummer Vinson Syndrome has been identified as a risk factor for squamous cell carcinoma of upper digestive tract in 3-15% of patients, most commonly among women between 15 to 50 years of age at the post cricoid region.⁵

Our 21 year old female patient presented with complaints of shortness of breath, and fatigue for the last 20 days with a past history of dysphagia to solid foods since childhood. In Chanakya et al study, a 38-year-old female, came with complaints of chest pain since one hour to the emergency

department with past history of dysphagia since one year, whereas in Bentoor et al study, a 25 year old Male complained of generalized weakness, shortness of breath since 15 days and dysphagia since 1.5 years.^{2,4} The female patient in our case is a known case of hypothyroidism and is on regular medication. Few authors suggest thyroid profile to rule out Hypothyroidism, as thyroid hormone plays a role in hemoglobin synthesis and hence may lead to anemia⁵.

Our patient had signs of conjunctival pallor and koilonychia on examination which are remarkable of IDA raising suspicion for further evaluation. Further hematological profile, revealed typical IDA (Hb:3.9gm/dl, ferritin levels:5.9ng/dl). Correspondingly, in Tomoitsu Tahara et al study, a 47-year-old female had hemoglobin levels of 3.8g/dl, ferritin levels of 2⁶. In another study by RM Hoofman et al, a 27-year-old female had Hb levels of 5.6g/dl⁷. For the further assessment of the cause of IDA, stool for occult blood and parasites was sent and was found to be negative. Upper Gastrointestinal Endoscopy revealed post cricoid webs and no negotiation of the endoscope further which was similarly seen in Bentoor et al study which could be the cause of her dysphagia to solid foods⁴. She was managed conservatively with parenteral iron supplementation. Upon gastroenterologist advise, esophageal dilatation was done, and there was a significant improvement in the symptoms. In contrast, in Chanakya et al study, patient was only kept on parenteral iron supplementation for a month with no dilatation of the esophagus, the symptoms of dysphagia decreased gradually. Though similarly, patient in Ajay et al study was treated with parenteral iron and esophageal dilatation^{1,2}.

As anemia and dysphagia can be effectively treated with iron supplementation it is considered that the general prognosis of this condition is good. However, prognosis deteriorates if associated with any malignancy. A surveillance upper GI endoscopy is required yearly.

Conclusion

PVS or Patterson Kelly syndrome is a rare syndrome with features of IDA, dysphagia, esophageal webs. It can be one of the differential diagnosis of dysphagia. Managing the esophageal webs of PVS and repleting iron stores at an early stage is crucial as the webs can progress to esophageal or pharyngeal squamous cell carcinoma.

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