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CASE REPORT OPEN ACCESS

Plummer-Vinson Syndrome: A Rare Case Report

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ABSTRACT

Plummer-Vinson syndrome, also called Paterson-Kelly syndrome, is characterized by the classic triad of dysphagia, iron-deficiency anemia, and esophageal webs. This syndrome is associated with an increased incidence of post-cricoid carcinoma and for surveillance, an endoscopy is recommended under general anesthesia. This was a case of 74-year-old woman with Plummer-Vinson Syndrome who was successfully treated with Savary-Gilliard (SG) dilation or Esophageal dilation. The patient had a long-standing clinical history of iron deficiency anemia with slow progression of dysphagia of solid food from 10 years and glossoepiglotic fold, aryepiglottic fold bilaterally arytenoid on the left & right side and had experienced difficulty in swallowing for the past 10 years. Along with this patient is having abdominal pain radiating to the epigastrium region and fever with chills & rigors. An endoscopy examination was conducted under general anesthesia and revealed the esophageal web at the level of the cervical esophagus. Laboratory data investigation shows an RBC count of 2.09 million/cumm, haemoglobin of 6.6 gm/dl%, and serum iron of 7µg/dl. The patient was prescribed Inj. Orofer-XT 100 mg intravenous iron sucrose supplement daily for 15 days, Inj. Pantoprazole 40 mg daily for 1 week, Inj. Tramadol 1amp whenever required and Syp.Sucralfate 15 ml 30 minutes before food. Her anemia condition was improved but dysphagia did not improve. To treat dysphagia the Savary-Gilliard (SG) dilation was done under fluoroscopy by endoscopically a single session was performed that serially increased the diameter by disrupting the web without any complication. After SG-dilation, the patient's dysphagia resolved shortly after the treatment.

Introduction

Plummer Vinson Syndrome (PVS), also called Paterson-Kelly syndrome, is characterized by the classic triad of dysphagia, iron-deficiency anemia and esophageal webs. The exact epidemiology of this syndrome is unknown, but it has become extremely rare. The condition is becoming less common in developed countries, but it is becoming more common in developing countries, especially in Asia. Iron deficiency is relatively common in African countries, but this syndrome is very rare [1]. Improvements in the nutritional status in the nations where Plummer-Vinson syndrome was previously reported have been proposed as the cause of the decline in the syndrome's prevalence [2].

It is more common in middle-aged women and usually affects perimenopausal women. It is linked to a higher risk of squamous cell carcinoma of the pharynx and proximal esophagus. It is referred to as Paterson-Brown-Kelly syndrome in the United Kingdom. The two British laryngologists who published their research in 1919, Adam Brown-Kelly (1865–1941) and Donald Ross Paterson (1863–1939), were honoured by this name [3,4]. Autoimmunity, genetic predisposition and dietary and iron deficits are some of the hypothesized etiopathogenic processes [5]. When iron deficiency repeatedly damages epithelia, it causes mucosal atrophy and pharyngeal muscle deterioration, which eventually results in the formation of esophageal webs. The esophageal web is asymmetrically linked to the anterior esophageal wall and is located beneath the cricopharyngeal muscle. Squamous epithelia make up the thin mucous membrane known as the esophageal web [6,7].

The widely accepted theory of iron insufficiency is still debatable. Previous studies have linked iron deficiency to the development of esophageal webs and dysphagia in people who are prone to it. Depletion of iron-dependent oxidative enzymes can lead to atrophy of the esophagus mucosa, the production of webs as an epithelial consequence and myasthenic alterations in the swallowing mechanism's of muscles. Plummer-Vinson syndrome has an enigmatic cause. Nutrient deficiencies, or the absence of specific nutrients, may have an impact, as well as genetic variables. It is an uncommon condition that has been connected to throat and esophageal malignancies. It affects women more frequently. The

tongue and oral mucosa of patients with Plummer-Vinson syndrome frequently experience burning and the dorsum of the tongue becomes smooth, glossy and red due to lingual papillae atrophy. Atrophic glossitis, angular cheilitis, koilonychia (abnormally thin nails, also called spoon nails), splenomegaly (an enlarged spleen), dysphagia, pain, weakness, odynophagia (painful swallowing) and upper esophageal webs (post cricoid region - contrasts with Schatzki rings found at the lower end of the Esophagus) are among the symptoms. The management mainly includes nutritional supplementation (iron supplement) and mechanical dilation or bougienage. By considering the few studies that have compared treatment modalities, prospective studies using mechanical dilation of esophageal results that 94% of patients had a complete response after the first session [8]. Along with a brief review of PVS, this case report of PVS emphasizes the need for the management or treatment and provides information among healthcare professionals as well as patients.

Case Report

A 74-year-old woman was admitted to the gastroenterology ward of the tertiary care hospital of Rajahmundry with a clinical history of iron deficiency anemia with slow progression of dysphagia of solid food from 10 years and she had a glossoepiglotic fold, aryepiglottic fold bilaterally arytenoid on the left and right side and had experienced difficulty in swallowing for the past 10 years. Along with this patient was having abdominal pain radiating to the epigastrium region and fever with chills & rigors.

Observation

Under general physical examination, blood pressure was 120/80mmHg, pulse rate was 76bpm, respiratory rate was 17/min, spO2 level was 99% and GRBS was 122mg/dl. On systemic examination, everything is found to be normal. The laboratory investigations showed the values of hemoglobin (6.6gm/dl), RBC (2.09 million/cumm), PCV (20.2%), WBC (2200 cells/cumm), reticulocyte count (1.2%) and serum iron (7 µg/dl). Along with this lipoprotein level, liver function test and renal function tests were found to be normal. An endoscopy examination was conducted under general anesthesia and revealed the esophageal web at the level of the cervical esophagus (Figure-1). By considering the general physical examination, laboratory investigation and endoscopic report the patient is diagnosed with Plummer Vinson Syndrome (PVS) and a standard form of treatment, the patient was prescribed with Inj. Orofer-XT 100 mg intravenous iron sucrose supplement daily for 15 days, Tab. Pantoprazole 40 mg daily for 1 week, Inj. Tramadol 1amp whenever required and Syp. Sucralfate 15 ml (30 minutes before food). Her anemia condition was improved but, dysphagia did not improve. To treat dysphagia the Savary Gilliard dilation (SG dilation) was done under fluoroscopy by endoscopically a single session was performed that serially increased the diameter by disrupting the web without any complication. After SG-dilation, the patient's dysphagia resolved shortly after the treatment. With this treatment, a significant improvement was seen in the patient.

The patient was admitted to the hospital for 15 days, after that the patient was discharged with the medication Tab. Oroferxt 1tab/day (100 mg ferrous ascorbate + 1.5 mg folic acid), Tab. Pantoprazole 40mg (an empty stomach), Tab. Ultracet (whenever required for the pain), Syp. Sucralfate 15 ml thrice a day (30 minutes before food) and Syp.Looz 10 ml 1 hour (before bed at night). The patient was suggested to come for follow up after 10 days.

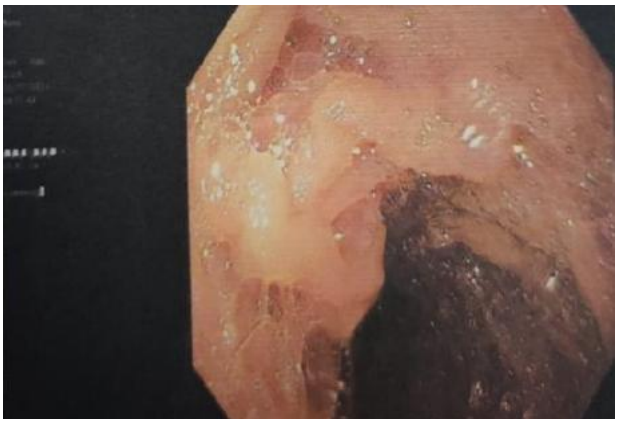

Figure 1: Before SG-Dilation Therapy

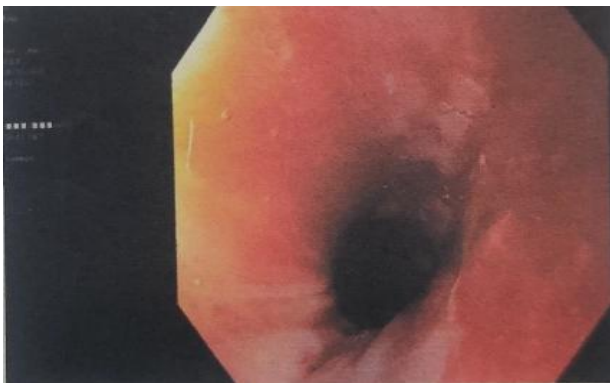

Figure 2: After SG-Dilation Therapy

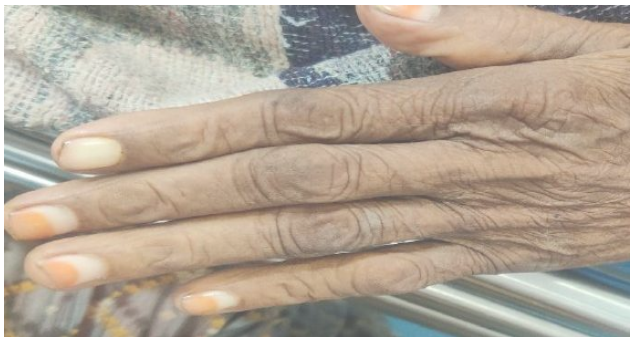

Figure 3: Koilonychia is not seen in the finger

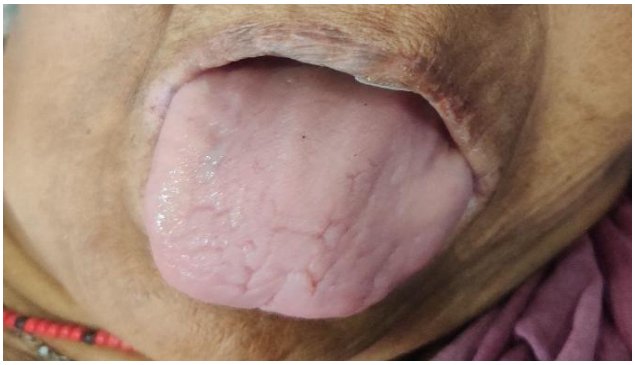

Figure 4: Angular Cheilitis and smooth tongue with the loss of the normal tongue papillae

Discussion

Plummer-Vinson Syndrome presents as a classical triad of iron deficiency anaemia (weakness, pallor, fatigue, tachycardia), post-cricoid dysphagia (painless & intermittent or progressive over years, limited to solids and sometimes associated with weight loss) and esophageal webs (at the level of the cervical esophagus). A 74-year-old woman was admitted in the gastroenterology ward with a clinical history of iron deficiency anemia with slow progression of dysphagia of solid food from 10 years and she had a glossoepiglotic fold, aryepiglottic fold bilaterally arytenoid on the left and right side and had experienced difficulty in swallowing for the past 10 years. On general physical examination and systemic examination, the patient was found to be normal. But laboratory investigations shown low level of hemoglobin, packed cell volume, white blood cells, serum iron and reticulocyte count. The patient was referred for endoscopy and it was found that esophageal web was found at the level of cervical esophagus. By considering subjective and objective evidence the final diagnosis is made as Plummer Vinson Syndrome. The treatment given to the patient was injection Orofer XT to address the iron deficiency associated with the syndrome, Inj. Tramadol for abdominal pain management, Tab. Pantoprazole for reducing gastric acid secretion that may contribute to overall management of PVS, syrup sucralfate for treating the ulcers by its mucosal protective effect. By this treatment her anaemic condition was improved much, but dysphagia condition was not subsided. For this, mechanical dilation was done to stretch or dilate the narrowed part of esophagus i.e., 1st session of SG dilation was done and found significant improvement in patient.

Conclusion

A 74-year-old woman diagnosed with Plummer Vinson Syndrome was treated with iron supplementation to treat iron deficiency anemia and for dysphagia, Savary Gilliard dilation (SG-Dilation) was effective in treating the esophageal web condition. It has been suggested that iron deficiency anemia with dysphagia in PVS is usually improved by iron supplements but in several cases in which the dysphagia did not respond with iron supplemental therapy the patient ultimately requires mechanic dilatation (SG dilation or bougienage). In this case, iron deficiency anaemia condition improved but the dysphagia condition did not subside so that SG dilation was done under fluoroscopy by endoscopically a single session was performed that serially increased the diameter by disrupting the web without any complication. After SG-dilation, the patient's dysphagia resolved shortly after the treatment.

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Conflict of interest

All authors declare that there is no conflict of interest.

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