CASE REPORT

Plummer-Vinson Syndrome: A Case Report

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Abstract

Plummer-Vinson syndrome (PVS), also termed as sideropenic dysphagia or Paterson-Brown-Kelly syndrome, is an uncommon medical disorder characterised by three things: iron deficiency anaemia, dysphagia, and esophageal web(s), more often seen in middle-aged females. We present a case of a 45-year-old married lady who came to the medical outpatient department at Abbasi Shaheed Hospital with food swallowing difficulty, fatigue, and generalised body weakness for the past 7 months. Her hematological workup revealed microcytic hypochromic anaemia and barium studies showed smooth circumferential narrowing of the cervical esophagus at the level of the C5-C6 vertebrae (post-cricoid esophageal web). PVS was diagnosed. Endoscopic dilatation or balloon dilatation was advised to treat her dysphagia but was refused by the patient. Her iron deficiency anaemia was corrected with an intravenous iron sucrose supplementation. She was encouraged to eat an iron-rich, semi-solid diet. When she came for a follow-up after six weeks of oral iron supplementation, her condition was satisfactory, and dysphagia had improved.

Keywords: *Dysphagia, anemia, Esophageal webs, Plummer-Vinson syndrome, Paterson-Brown-Kelly syndrome.*

INTRODUCTION

Plummer-Vinson syndrome (PVS) is a rare medical disorder clinically manifested as dysphagia, iron-deficient anaemia, and cervical esophageal web(s). If left untreated, these webs may raise the risk of esophageal squamous cell carcinoma in these patients. Patients may exhibit other signs and symptoms of anaemia, like pallor on the face or conjunctiva, breathlessness, fatigue, an atrophic smooth tongue, ridges on nails (Koilonychias), angular cheilitis, *etc.* [1]. PVS primarily impacts middle-aged women. It has rarely been noted in males [2]. PVS is diagnosed through a thorough medical history, general clinical examination, haematological analysis, barium studies, or upper gastrointestinal endoscopy [3].

Even though this syndrome is extremely uncommon these days, it is still crucial to recognize it to lower the chance of esophageal cancer [4].

CASE REPORT

A 45-year-old married lady with a known case of hypertension visited the medical outpatient department at Abbasi Shaheed Hospital complaining about trouble swallowing food, fatigue, and generalised body weakness for the past 7 months, for which she acquired admitted to the medical ward.

As per the patient, she was alright 7 months before, when she began to have difficulty in swallowing solid meals and got easily fatigued while doing her normal routine work. She had noted a gradual decline in her health over the past few months. Her dysphagia was associated with generalised body weakness, reduced appetite, undocumented weight loss, dizziness, and palpitations. Her dietary intake was unsatisfactory. She denied any history of breathing difficulty, dark-colored stools, blood loss, pica, abdominal pain, fever, or cough. Her gynaecological history was insignificant. She denied any history of malignancy or tuberculosis in her family.

On a general physical examination, the patient was malnourished and ill-looking, well-oriented to time, place, and person. Regarding her vitals, her blood pressure was 120/70 mmHg, her pulse was 102 beats per minute, her respiratory rate was 18 breaths per minute, and her SpO2 was 95%. The patient had severely pale conjunctiva, ridging on her nails (Koilonychias), and angular cheilitis. Signs of malnutrition like interossei guttering and muscle wasting were present. Her abdominal,

Table 1: Laboratory results.

Hematological Examination	Patients Lab Results	Normal Range
Haemoglobin	7.5 g/dl	13-17 g/dl
Haematocrit	19%	36-48%
MCV	68 fl	80-99 fl
MCH concentration	21 g/dl	32-36 g/dl
Serum Iron	38 µg/dl	50-150 µg/dl
Serum TIBC	490 µg/dl	250-450 µg/dl
Serum ferritin level	16 ng/ml	24-307 ng/ml
TLC	6300/µl	4000-11,000/µl
Platelet count	457,000/µl	157,000-371,000/µl

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cardiovascular, and respiratory examinations were unremarkable. Her laboratory investigations were done shown in **Table 1**. Peripheral blood film revealed microcytic hypochromic anaemia. Her iron profile showed low serum iron and ferritin levels with high TIBC. An initial diagnosis of iron deficiency anaemia was made. The stool was negative for occult blood to rule out any blood loss from the gastrointestinal tract. Hb electrophoresis, renal function test, liver function test, ultrasound of the whole abdomen, and urine analysis were all within normal limits.

A suspected diagnosis of PVS was made because of iron deficiency anemia and dysphagia. Barium studies were advised, which showed smooth circumferential constriction of the upper/cervical esophagus at the C5-C6 vertebral level as shown in **Fig. (1)**. A mild holdup of contrast was noted at the site of narrowing. According to the radiologist, the circumferential narrowing at C5-C6 represents post post-cricoid esophageal web. Her PVS diagnosis was confirmed by a triad of iron deficient anemia, dysphagia, and esophageal web(s). Endoscopic dilatation/balloon dilatation was refused by the patient. Her iron deficit was calculated and an appropriate dose of an intravenous iron sucrose along with some appetite stimulants was given. Iron-rich semi-solid diet and oral iron supplements were advised. After six weeks, she came for a follow-up and her condition had improved.

DISCUSSION

Epidemiology

Exact data on the syndrome's incidence and prevalence are not available. Plummer-Vinson syndrome appeared to be frequent in Caucasians of Northern countries in the first half of the twentieth century, notably among middle-aged women but some pediatric and adolescent cases, however, have been reported [1]. It is now extremely rare. The quick decline in the disease's prevalence corresponds to improved nutritional status and the removal of widespread iron deficiency in regions where the syndrome was previously characterized [2].

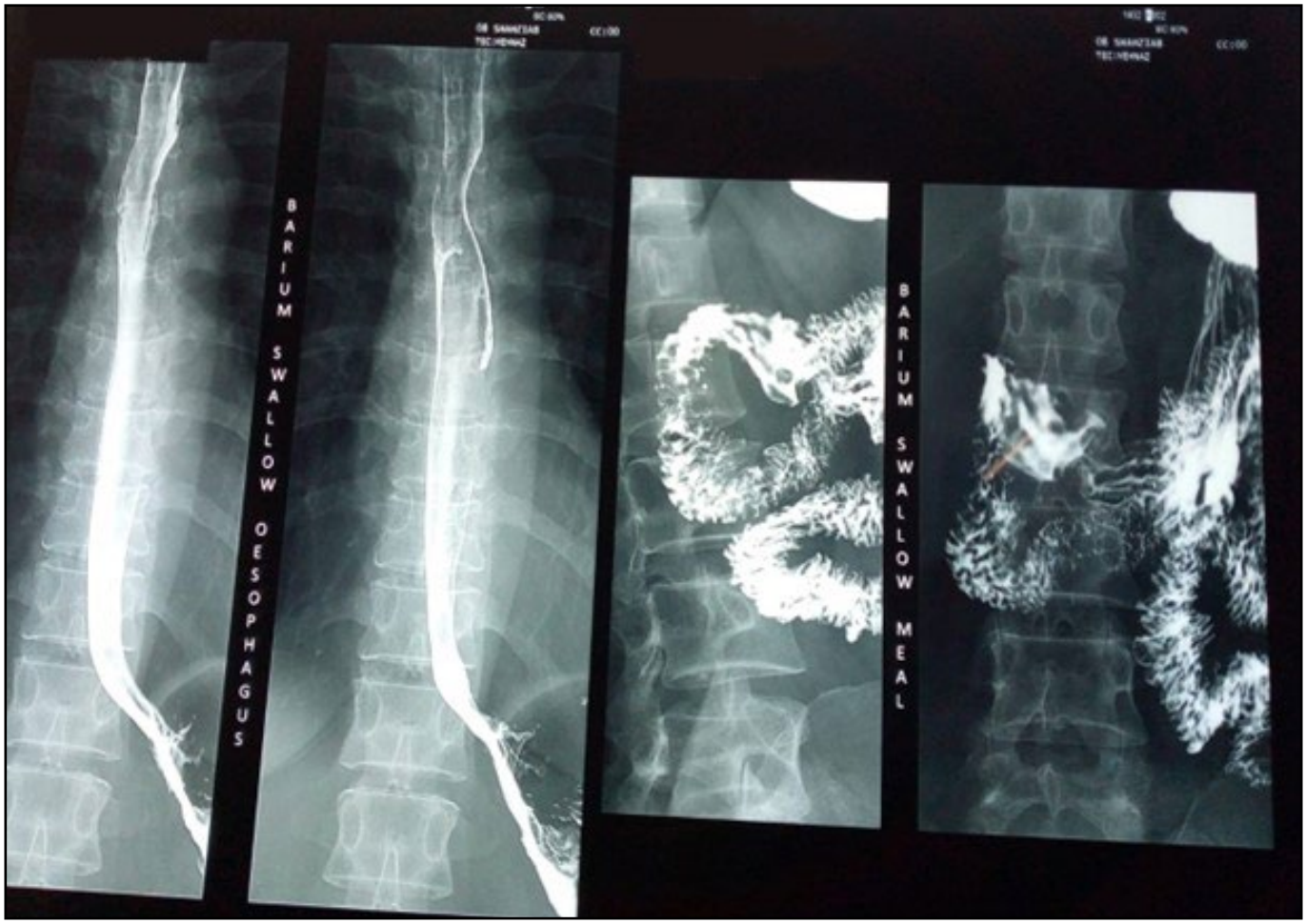

Fig. (1): Barium studies showed smooth circumferential narrowing of the cervical esophagus at C5-C6 vertebrae (Post cricoid esophageal web).

The cross-sectional study was conducted at Bahawalpur Victoria Hospital, where 164 male and female patients with dysphagia were reported from April 1st, 2020 to March 31, 2021, with 11.0 patients identified with PVS. Age does not correlate with the duration of dysphagia ($p>0.05$), however, gender shows an association with PVS ($p<0.05$) [5]. Recently, a few instances of PVS in male patients have been published, raising the possibility that PVS is widespread in both males and females [6]. However many studies show that PVS has an unusually high female-to-male ratio of 4:1 [2].

One more study in Pakistan is From June 2009 to June 2015, This cross-sectional study was carried out in the Department of Otolaryngology at Khyber Teaching Hospital in Peshawar, Pakistan. A total of 423 patients were chosen as the sample size, with 21 patients having PVS in dysphagia, one of which was male and the other 20 being female [7].

Our case report also describes a middle-aged female patient who presented with PVS clinical characteristics.

Etiopathogenesis

PVS pathogenesis is still mainly unknown. However, iron insufficiency is the most likely cause of PVS. Iron deficiency results in the depletion of iron-dependent enzymes [8]. A lack of iron-dependent oxidative enzymes leads the pharyngeal muscles to degrade slowly, leading to mucosal atrophy, web(s) formation, and eventually, the development of upper esophageal cancer [9]. Physical manifestations of tissue iron shortage consist of pale conjunctiva, angular cheilitis, and koilonychia, all of which have been seen in our patient.

Laboratory Test(s)

Although the specific cause and pathogenesis of PVS are unknown, iron and other dietary deficiencies, genetic susceptibility, and autoimmune disorders have all been linked to the creation of the webs. Hematological examination often demonstrates iron deficiency anemia with lower hemoglobin, hematocrit, MCV, and serum ferritin levels and higher total iron binding capacity. A few publications recommend a thyroid profile to rule out hypothyroidism because thyroid hormones are involved in hemoglobin production and can cause anemia [10]. In this case report, hematological tests revealed that our patient had severe iron-deficient anemia.

Radiographic Examination

Endoscopy or radiography procedures can detect esophageal webs and strictures. However, radiography is more appropriate because endoscopy can occasionally miss the point of benign stricture and does not detect the majority of motility abnormalities [11]. The most sensitive test for detecting esophageal webs is the barium swallow test. Although barium sulfate is an inert substance that creates a strong contrast, a thick paste should be utilized with rapid exposure after eating for optimal visualization [10]. In our investigation, our patient's barium swallow test revealed smooth circumferential constriction of the upper/cervical esophagus at the C5-C6 vertebral level.

Diagnosis

PVS is diagnosed when a patient with post-cricoid dysphagia has iron deficiency anaemia and one or more esophageal webs. As a result, a complete history, general clinical examination, haematological test (anaemia profile), and radiographic exam (simple lateral X-ray of the neck and barium swallow test) are used to make the diagnosis. In rare cases, additional testing such as direct endoscopic examination, videofluoroscopy, and biopsy for histological evaluation may be required [1, 2]. Our patient was diagnosed with PVS after a haematological and radiographic evaluation; endoscopic dilatation was also recommended but denied by the patient.

Differential Diagnosis

Because dysphagia is a key clinical characteristic of Plummer-Vinson syndrome, any alternative causes of dysphagia, including malignancies, esophageal rings, or benign strictures must be considered. Diverticula, motility disorders such as achalasia, spastic motility disorders, diabetes mellitus, gastric reflux disease, scleroderma, and neuromuscular and skeletal muscle disorders are all causes of dysphagia [2].

Malignant Potential

PVS has been found as a contributory factor for developing squamous cell carcinoma of the upper aerodigestive tract in 3%-15% of patients, with the majority of cases occurring in women between the ages of 15 and 50, and virtually always occurring in the post-cricoid region [2]. The mechanism is that anaemia promotes epithelial atrophy and reduces the mucosa's healing capacity, allowing carcinogens and co-carcinogens to act strongly, exposing the entire mouth cavity and esophageal area to cancer [12]. A rare link of PVS with a base of tongue malignancies has been documented in the literature [13]. Hence, these individuals should be recommended to get proper follow-up care and upper gastrointestinal endoscopies to rule out any neoplastic alterations.

Treatment

It has been recommended that iron supplements can help with dysphagia caused by PVS [14]. There have also been reports of dysphagia that did not respond to iron therapy and needed endoscopic dilatation or incision [15]. Because our patient refused endoscopic dilatation/

balloon dilatation, iron therapy was administered, and this greatly improved her dysphagia. Furthermore, if iron deficiency reoccurs, esophageal webs may develop again, therefore, these patients must be closely monitored. PVS has also been related to an increased chance of upper gastrointestinal cancer. Endoscopic screening is also required due to the danger of cancer.

CONCLUSION

PVS is an uncommon medical condition that primarily affects middle-aged females. Iron supplementation shows improvements in anaemia and dysphagia. Some patients may need endoscopic esophageal web dilatation for the treatment of dysphagia. Early diagnosis is very important for a better prognosis and to reduce the risk of upper esophageal malignancy, as it is a premalignant condition.

CONSENT FOR PUBLICATION

Informed consent was taken from the patient for publication.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

ACKNOWLEDGEMENTS

Declared None.

AUTHOR'S CONTRIBUTION

This study was designed and developed by ML, SA, and AA. ML and SHN conducted their preliminary literature review. MAR and WY were in charge of data gathering, assembly, and patient evaluation. The manuscript was written by ML, SA, and AA. The last critical review and changes were completed by ML and MAR. On behalf of all other authors, ML is the corresponding author.

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