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Case Report

An elusive diagnosis: a case of Plummer-Vinson syndrome

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ABSTRACT

Plummer-Vinson syndrome (PVS), also known as sideropenic dysphagia or Paterson-Brown-Kelly syndrome, is a rare medical disorder characterised by triad of iron deficiency anaemia, dysphagia, and oesophageal webs, more often seen in middle-aged females. We present a case of a 37-year-old female who came with complaints of difficulty in swallowing food, fatigue, and generalised body weakness for the past 6 months. Her hematological workup revealed microcytic hypochromic anaemia and barium studies showed smooth concentric narrowing of the cervical oesophagus at the level of the C5-C6 vertebrae (post-cricoid oesophageal web). Hence a diagnosis of Plummer -Vinson syndrome was made. Endoscopic dilatation was done to treat her dysphagia. Her iron deficiency anaemia was corrected with 1-unit prbc transfusion and intravenous iron sucrose supplementation. She was encouraged to eat an iron-rich 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, Oesophageal 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 oesophageal web(s). If left untreated, these webs may raise the risk of oesophageal squamous cell carcinoma in these patients. Patients may show other signs and symptoms of anaemia, like pallor on the face or conjunctiva, breathlessness, fatigue, an atrophic smooth tongue, spooning on nails (Koilonychias), and angular cheilitis.[1]

PVS primarily affects 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 recognise it to lower the chance of oesophageal cancer.[4]

CASE REPORT

A 37-year-old female with no significant comorbidities presented with troubled swallowing, fatigability and on and off dizziness for the past 6 months. Dysphagia was insidious in onset gradually progressive in nature, predominantly to solid foods. Her dysphagia was associated with generalised weakness, reduced appetite, undocumented weight loss, dizziness, and palpitations. She denied any history of breathing difficulty, dark-coloured stools, blood loss, abdominal pain, fever, or cough. Her gynaecological history was insignificant. She denied any history of malignancy or tuberculosis in her family.

On examination

The patient was poorly nourished and moderately built, well-oriented to time, place, and person. 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 SpO_2 was 95%. The patient had severely pale

conjunctiva, spooning of her nails (Koilonychias), and angular cheilitis. Her abdominal, 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. A first diagnosis of iron deficiency anaemia was made. The stool was negative for occult blood to rule out any blood loss from the gastrointestinal tract, renal function test, liver function test, Hb electrophoresis, ultrasound of the whole abdomen, and urine analysis were all within normal limits.

Table 1: Lab parameters.

Hematological examination	Patients lab result	Normal range
Hemoglobin (g/dl)	5.9	12-18
Hematocrit (%)	21.7	36-47
MCV (fl)	59	82-92
MCHC (g/dl)	27.1	32-36
Serum iron (mcg/dl)	22	50-150
Serum ferretin	<1.50	Female: 22-322
TLC (cells/micro-litre)	6100	4000-11000
Platelet count (lakhs/Cumm)	3.68	1.5-4

A suspected diagnosis of PVS was made because of iron deficiency anemia and dysphagia. Barium studies were advised, which showed smooth concentric narrowing of the upper/cervical oesophagus at the C5-C6 vertebral level as shown in Figure 1.

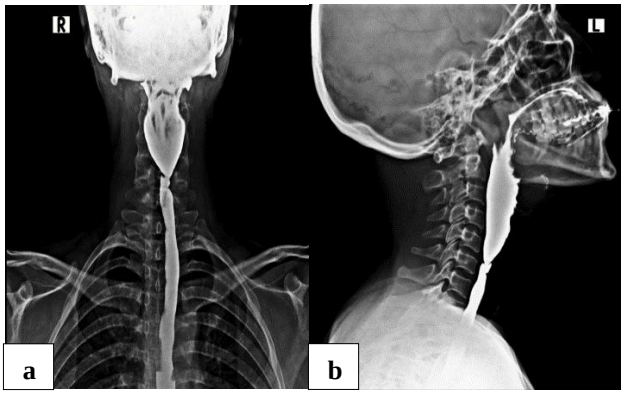

Figure 1 (a and b): Barium studies show smooth concentric narrowing of the upper/cervical oesophagus at the C5-C6 vertebral level, thin transverse filling defect was noted in the anterior aspect of the cervical oesophagus at the level of C6 vertebrae (post cricoid esophageal web).

A thin transverse filling defect was noted in the anterior aspect of the cervical oesophagus at the level of C6 vertebrae. According to the radiologist, they represent post-cricoid oesophageal web. Her PVS diagnosis was confirmed by a triad of iron deficient anemia, dysphagia, and oesophageal web(s). The patient underwent endoscopic dilatation after 1-unit PRBC transfusion. 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.

Diagnosis

Diagnosis was Plummer-Vinson syndrome.

Treatment

Endoscopic/balloon dilatation with parenteral and oral iron supplementation.

Follow-up

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 paediatric 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 characterised.[2] Many studies show that PVS has an unusually high female-to-male ratio of 4:1.[2]

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.[5] 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 oesophageal cancer.[6] 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.[7] 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.[8] The most sensitive test for detecting esophageal webs is the barium swallow test. Although barium sulphate is an inert substance that creates a strong contrast, a thick paste should be used with rapid exposure after eating for optimal visualization.[7] In our investigation, our patient's barium swallow test revealed smooth circumferential constriction of the upper/cervical oesophagus 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, video fluoroscopy, 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 done for 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.[9] A rare link of PVS with a base of tongue malignancies has been documented in the literature.[10] 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.[11] There have also been reports of dysphagia that did not respond to iron therapy and needed endoscopic dilatation or incision.[12] 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 oesophageal web dilatation for the treatment of dysphagia. Early diagnosis is very important for a better prognosis and to reduce the risk of upper oesophageal malignancy, as it is a premalignant condition.

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