CASE REPORT
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Case series of Plummer-Vinson syndrome from Ethiopia, Sub Saharan Africa

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Competing interests
The authors declare no conflicts of interest.

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Abbreviations
EGD, Esophagogastroduodenoscopy; CBC, Complete Blood Count; FNAC, Fine Needle Aspiration Cytology; Hb, Hemoglobin; MCH, Mean Corpuscular Hemoglobin; MCHC, Mean Corpuscular Hemoglobin Concentration; MCV, Mean Corpuscular Volume; OPD, outpatient department; PVS, Plummer-Vinson Syndrome; TFT, Thyroid Function Test; TIBC, Total Iron-Binding Capacity; TSH, Thyroid-Stimulating Hormone.

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Abstract
Plummer-Vinson syndrome (PVS) is a rare condition marked by postcricoid dysphagia, upper esophageal webs, and iron deficiency anemia. It has been associated with increased risk of hypopharyngeal and esophageal malignancies, necessitating early diagnosis. This case series reviews four patients who presented to Adera Medical and Surgical Center in Addis Ababa, Ethiopia, with PVS. All patients exhibited progressive dysphagia, anemia, and fatigue. Endoscopic evaluation revealed esophageal webs in the upper esophagus, and treatment included iron replacement therapy and esophageal dilation as needed. Hemoglobin levels improved in all cases, with three patients experiencing complete resolution of dysphagia, while one required a repeat dilation after 6 months for recurrence.

Keywords: Plummer-Vinson Syndrome (PVS); dysphagia; esophageal webs; iron deficiency anemia

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Introduction

Plummer-Vinson syndrome (PVS), also called Paterson-Brown-Kelly syndrome, is a rare entity whose main clinical features are upper esophageal web(s), dysphagia and iron deficiency anemia. This syndrome has been known since the beginning of the 20th century and is named after two American Physicians Henry Stanley Plummer and Porter Paisley Vinson. The term sideropenic dysphagia has also been used, because the syndrome can occur with iron deficiency (sideropenia), but it is not associated with anemia [1, 2].

Postulated etiopathogenic mechanisms include iron and nutritional deficiencies, genetic predisposition, and autoimmune factors, amongst others [1]. The depletion of iron-dependent oxidative enzymes may produce myasthenic changes in muscles involved in the swallowing mechanism, atrophy of the esophageal mucosa, and formation of webs as epithelial complications [3]. The improvement in dysphagia after iron therapy provides evidence for an association between iron deficiency and post-cricoid dysphagia [4]. PVS has also been viewed as autoimmune phenomenon since it has been associated with autoimmune condition, such as rheumatoid arthritis, pernicious anemia, celiac disease, and thyroiditis [5].

The typical age range at diagnosis is 40–70 years. It has been described in whites and is more frequently observed in women with good prognosis unless complicated by hypopharyngeal or esophageal carcinoma [6].

Plummer-Vinson syndrome is usually managed on out-patient basis with Iron replacement therapy to correct the anemia and to resolve most of the physical signs of deficiency. Dysphagia may improve with iron replacement alone, particularly in patients whose webs are not substantially obstructive. If the webs are more advanced, response to iron replacement alone is unlikely and thus is managed with mechanical dilation [7].

Despite a century since its first description, literature on PVS remains limited to isolated case reports. While PVS has become rare in developed regions like Europe and North America due to improved nutrition and iron supplementation, its prevalence has risen in developing countries, particularly in Asia. However, in Africa, where iron deficiency is widespread, PVS remains largely unrecognized, with very few documented cases, especially in sub-Saharan Africa possibly due to under-diagnosis because of the limitation of diagnostic and therapeutic means or under-reporting [8].

In this case series, we present four patients with PVS who visited our medical center with the complaint of dysphagia and anemia at different times over the past 11 months and were treated with endoscopic dilation and iron replacement. Endoscopic Savary-Gilliard dilators were used during endoscopic dilation after numbing the throat with xylocaine spray, with light sedation to enhance patient comfort without increasing complication risks. The successful rupture of the web was confirmed by the presence of a small amount of fresh blood at the site during endoscopy.

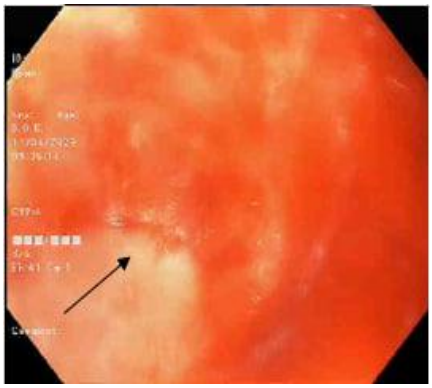
Case 1

A 41-year-old male presented to our outpatient department (OPD) at Adera Medical and Surgical center with a complaint of long standing and progressive painless dysphagia of 3 years which initially was for solid foods and later became for liquid meals. He also reported associated unspecified weight loss, easy fatigability, exertional dyspnea, blurring of vision and dizziness.

He had stable vital signs and pale conjunctiva otherwise no abnormality was detected on physical examination. On investigation, complete blood count (CBC) revealed hemoglobin (Hb) of 7.9 g/dl, mean corpuscular volume (MCV) of 55.9, mean corpuscular hemoglobin (MCH) of 18.5 and mean corpuscular hemoglobin concentration (MCHC) of 30.9. Liver function test, renal function test, erythrocyte sedimentation rate, and abdominal ultrasound were all normal.

Esophagogastroduodenoscopy (EGD) showed upper esophageal (20 cm from incisors teeth) luminal stenosis with visible semicircular web. Scope was passed with moderate pressure and the membrane of the web was ruptured with no significant bleeding or complication (Figure 1). The rest of the esophagus, stomach and duodenum were all normal. The patient was given oral ferrous sulfate 325 (65 Fe) mg. On his follow up visit after a month, the dysphagia improved, gained 3 kg of weight and Hb normalized. There were no complications, relapses, or need for further treatment following his initial presentation, which was 11 months ago.

(A)



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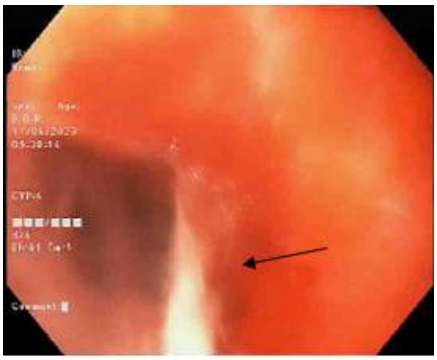


Figure 1 Endoscopy showing upper esophageal luminal stenosis with visible semicircular web 20 cm from incisors teeth (A) Post dilation membranous web (B)

Case 2

A 29-year-old female presented to our OPD at Adera Medical and Surgical center with a complaint of long-standing difficulty of swallowing for solid food associated with anterior neck pain. She has also easy fatigue associated with light headedness, palpitations, occasional epigastric pain and bloating and insomnia.

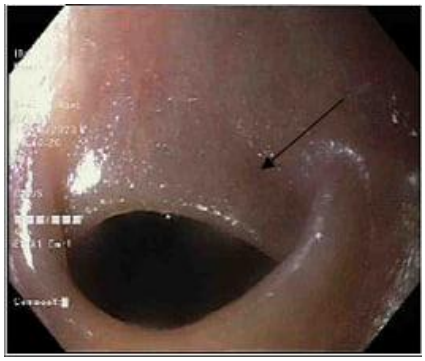
Upon presentation, she had stable vital signs with mild pallor. Otherwise no abnormality was detected on physical examination.

Laboratory investigations revealed Hb of 8.4 g/dl, MCV 57, MCH 15 and MCHC 27, serum Iron 18 µg/dL, TIBC 420 µg/dL transferrin saturation 4.29% serum ferritin < 1.98. Her thyroid function test (TFT) results showed TSH-19.34, T3-1.03, T4-73.74. Liver function test, renal function test, erythrocyte sedimentation rate and abdominal ultrasound were all unremarkable. Neck ultrasound reported diffuse goiter with enlarged thyroid lobes (RT = 2.2 × 1.7 cm; LT = 2.1 × 1.6 cm) and isthmus of which a fine needle aspiration cytology (FNAC) showed lymphocytic thyroiditis.

EGD revealed proximal esophageal web noted with sever stenosis at 15 cm and serial Savary-Gilliard wire guide dilation was done smoothly up to 12 mm and opened up with web tear and minor mucosal bleeds. No complication was encountered (Figure 2).

With the diagnosis of PVS and hypothyroidism the patient was treated with oral ferrous sulfate 325 (65 Fe) mg and thyroxine and improved with normal Hb and TFTs and remained symptom free for the past 10 months.

(A)



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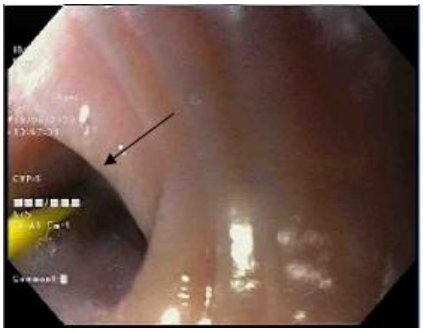

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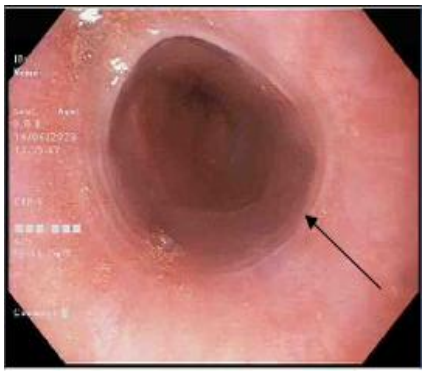


Figure 2 Endoscopy showing (A) upper esophageal luminal stenosis with visible semicircular web 15 cm from incisors teeth (B) wire guide dilation (C) Post dilation

Case 3

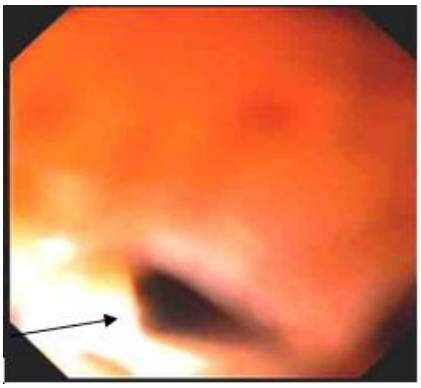
A 48-year-old female presented to our OPD at Adera Medical and Surgical center with a complaint of painless progressive dysphagia of seven months which initially was for solid foods and later became for liquid meals also. Associated to this, she has postprandial bloating, and reflux with weight loss of 3 kg.

Upon presentation, she had stable vital signs and no abnormality was detected on physical examination. On investigation, CBC revealed Hb of 8.1 g/dL with MCV of 55.2, MCH 16.8 and MCHC 30.5. Liver function test, renal function test, erythrocyte sedimentation rate, Lipid profile, and abdominal ultrasound were all unremarkable.

EGD revealed multiple semi-lunar webs in the proximal esophagus at the level of upper esophageal sphincter, 15cm from the incisors, with marked luminal narrowing precluding scope passage (Figure 3).

With the diagnosis of PVS, Iron replacement therapy was started with ferrous sulfate 325 (65 Fe) mg and her symptoms and the patient remained asymptomatic with weight gain of 2 kgs and Hb improvement to 12.9 g/dl on her follow up visit after 1 month. There were no complications, relapses, or need for further treatment over the last 8 months.

(A)



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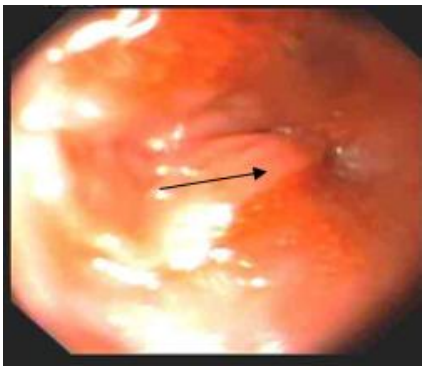


Figure 3 Endoscopy showing a semi-lunar web in the proximal esophagus at the level of upper esophageal sphincter (A), with marked luminal narrowing (B)

Case 4

A 27-year-old female presented to our OPD at Adera Medical and Surgical center with a complaint of progressive painless dysphagia with intermittent choking episodes of 5 years. The dysphagia was initially for solid foods and later for liquid. She also had associated postprandial bloating, and reflux, blurring of vision and dizziness. She also complained heavy menses.

Physical examination revealed normal vital signs and pale conjunctiva otherwise unremarkable. Her blood work up revealed Hb of 8.4 g/dl with MCV of 66.9, MCH 21.1 and MCHC 28.7 and peripheral morphology reported hypochromic microcytic anemia.

Barium swallow study showed ring like transverse constriction of the mid part of cervical esophagus circumferential cervical esophageal web (Figure 4) which was confirmed by follow up EGD showing a proximal web with sever stenosis noted at 15 cm from the incisors and serial Savary-Gilliard wire guided dilation done smoothly (5–10 mm) and opened up with mucosal and membrane tear and minor bleeding (Figure 5).

With the diagnosis of PVS, Iron replacement therapy with ferrous sulfate 325 (65 Fe) mg was started and her symptoms improved and Hb was 13.5 during her follow up visit after 1 month. Six months after her initial presentation she complained of dysphagia. Her blood works revealed Hb of 8.0 g/dL and Abdominopelvic Ultrasound reported a 4 × 5 cm intramural myoma. A repeat EGD showed esophageal webs after which second session of dilation was done. The patient was then supplemented with iron and linked to a gynecologist for further management.

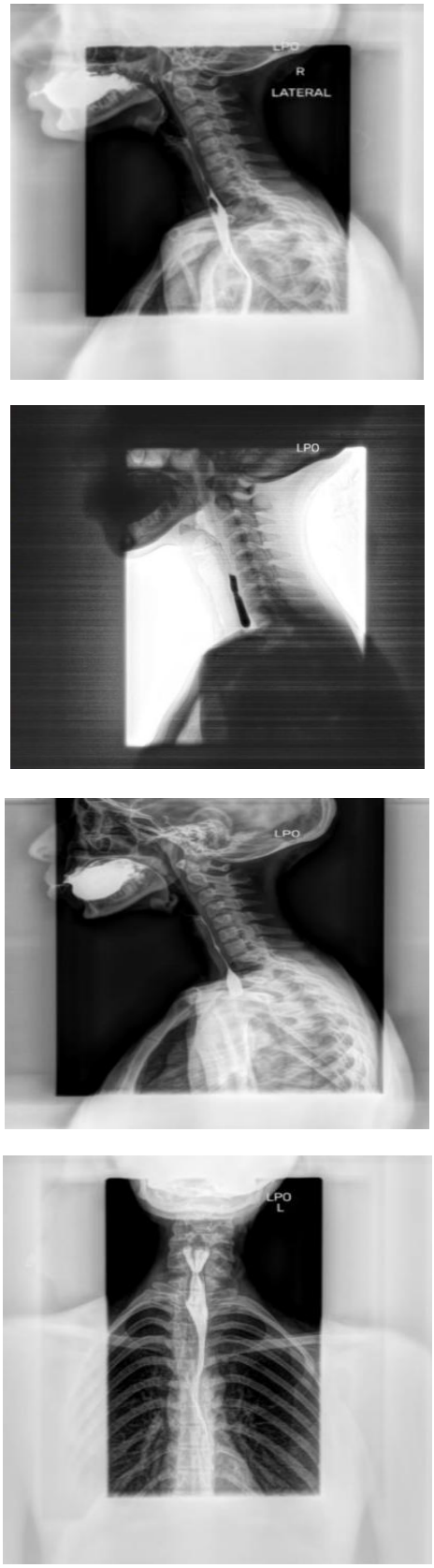
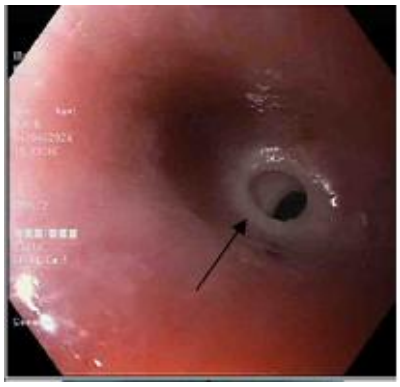


Figure 4 Barium swallow study images showing a circumferential esophageal web

(A)



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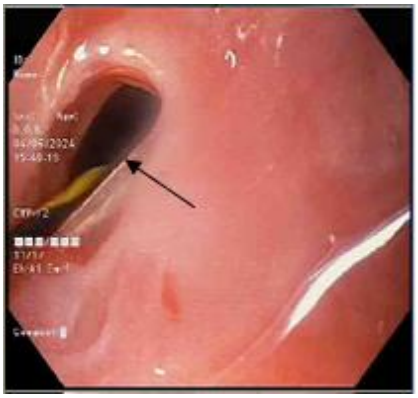

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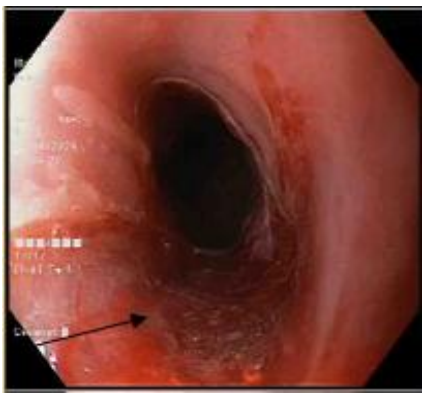

Figure 5 Endoscopy showing a semi-lunar web in the proximal esophagus at the level of upper esophageal sphincter with marked luminal narrowing (A), wire guide dilation (B), post dilation (C)

Discussion

PVS is a rare syndrome, and data on its prevalence in modern times is limited. It is believed to be significantly more common in females; however, recent reports of PVS in male patients suggest that the condition may be common in both genders than previously thought [9, 10]. In line with this, the first case in our series involves a 41-year-old male presenting with classic features of PVS. Similarly, a case series of four patients from Ethiopia also reported two male patients [11].

The pathogenesis of PVS remains largely unknown. However, the most probable mechanism is iron deficiency which leads to rapid loss of iron-dependent enzymes. Loss of these enzymes cause web formation and eventually leads to cancer development of the upper gastrointestinal tract [12]. Additionally, PVS has been reported in

association with autoimmune conditions like thyroiditis, rheumatoid arthritis, and Crohn's disease, raising the possibility that immune dysregulation may be involved in its pathogenesis, although this has yet to be proven [9, 13]. In our series, the second case involved a patient with sideropenic dysphagia who was also diagnosed with lymphocytic thyroiditis. Similarly, in a retrospective study from Senegal, one out of 50 patients with PVS was found to have hyperthyroidism [14].

Most patients with PVS are initially asymptomatic. Similar to our cases, dysphagia is painless and slowly evolving, starting with solid foods and difficulty swallowing liquids after years of initial onset [10]. Dysphagia becomes symptomatic only when the luminal diameter in the region of the esophageal web becomes less than 12 mm. It is divided into two classes, grade I (occasional dysphagia on taking solids) or grade II (able to swallow only semi-solid diet) [9].

The epithelial layer of the upper alimentary tract is especially susceptible to iron deficiency because of its high cell turnover. Studies suggest that dysphagia associated with PVS is improved by iron supplements while there have also been several cases in which the dysphagia did not respond to iron therapy and ultimately required endoscopic dilation or incision [4]. Determining the underlying cause of iron deficiency is also crucial, as it may require targeted treatment, such as in cases of celiac disease [5].

Those with advanced and long-standing dysphagia typically require mechanical dilation. Commonly used techniques include endoscopic balloon dilatation or savary-Gilliard dilators. Esophageal webs may relapse if iron deficiency recurs; therefore, careful follow up is mandatory for such patients [13]. A Tunisian study involving 23 patients with PVS reported a recurrence of dysphagia in 5 patients, with a median time of 26.6 months (ranging from 2 to 60 months) after initial treatment with iron supplementation and endoscopic dilation. These recurrences required a second session of dilation, and one patient, who had refractory anemia, required four sessions [8]. In our series, the fourth patient experienced a recurrence of dysphagia 6 months after her initial dilation due to an esophageal web caused by anemia secondary to a gynecological condition. She underwent a repeat endoscopic dilation.

The prognosis of PVS is generally good for symptom control, but it is considered a precancerous condition with an increased risk of developing squamous cell carcinoma of the hypopharynx or upper esophagus. Studies report post-cricoid carcinoma in 4% to 16% of PVS patients. The exact cause of this transformation is unknown but may be linked to persistent iron deficiency leading to irreversible atrophic mucosal changes, which may eventually result in malignant degeneration for some patients. Although there is no established guideline for follow up of patients with PVS, it is recommended that patients undergo annual reassessment for the recurrence of dysphagia or other early signs of cancer [9]. In our case all the patients were informed on the need for long-term monitoring and to look out for symptoms such as anorexia, weight loss, odynophagia, voice changes, or nasal regurgitation of food or liquids as well as the importance of follow-up visits to detect any recurrence of esophageal webs or malignancy.

There are various steps in the natural history of the disease in patients with PVS where multidisciplinary care and communication is essential. Coordination of care between the primary care providers and gastroenterologists is necessary. In upper endoscopic evaluation, gastroenterologists should be informed of the suspicion of esophageal webs as they are easily missed and sometimes, just passage of the endoscope itself can cause rupture of the esophageal web [15].

Conclusion

PVS remains a rare but important cause of dysphagia among patients with iron deficiency anemia. In regions such as Sub-Saharan Africa, including Ethiopia, where iron deficiency is prevalent, clinicians should maintain a high index of suspicion for PVS in patients presenting with dysphagia and anemia. Early diagnosis and treatment with iron supplementation and endoscopic dilation can lead to significant symptom relief and prevent potential complications. Regular follow-up and cancer surveillance are also critical in managing the long-term risks associated with this syndrome.

Consent for publication

Written informed consent was obtained from the patients for publication of this case series and the accompanying images.

Availability of data and materials

The datasets used on this series are available from the principal investigator up on reasonable request.

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