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CASE SERIES

A REPORT OF FOUR ETHIOPIAN PATIENTS WITH PLUMMER –VINSON SYNDROME

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ABSTRACT

Plummer–Vinson syndrome (PVS) is a rare condition characterized by a triad of dysphagia, iron deficiency anemia, and upper esophageal web. The post-cricoid web is associated with increased risk of development of squamous cell carcinoma of the esophagus. In this presentation, we report four Ethiopian patients with Plummer–Vinson syndrome. The patients presented with dysphagia, iron deficiency anemia and typical endoscopic features of the syndrome; the patients were treated with endoscopic dilation and oral iron replacement.
Key words: Dysphagia, anemia, upper esophageal web, Plummer–Vinson syndrome

INTRODUCTION

Plummer-Vinson syndrome (PVS) is characterized by dysphagia, iron deficiency anemia and upper esophageal web(s) (1,2). The syndrome, known as PVS in the United States, is called Paterson-Brown Kelly syndrome in United Kingdom. It is also known as sideropenic dysphagia as it is associated with iron deficiency (sideropenia).

This syndrome has been known since the beginning of the 20th century after Stanley Plummer and Paisley Vinson reported a series of cases, which were further described by Ross Paterson and Brown Kelly. The syndrome has become a rare condition in the 21st century because of the improvement in nutritional status, advancement in medical tools, increased awareness and treatment of this condition and the underlying causes.

In this report, we present four cases with dysphagia and anemia. The patients came to Tikur Anbessa Specialized hospital at different times but within a period of two years. All patients were treated with endoscopic dilation and iron replacement.

Case 1: A 16-year-old female presented with long standing and progressive dysphagia of five years duration. The dysphagia is mainly to solid foods. She had no weight loss or abnormal menstrual bleeding. Clinical examination showed pale conjunctivae, atrophic tongue, angular cheilitis and spooning of the finger nails. Laboratory tests showed iron deficiency anemia: hemoglobin (Hg) =7.3 g/dl, mean corpuscular volume (MCV)=57.2 fL, mean corpuscular hemoglobin (MCH)=18.7, mean corpuscular hemoglobin concentration (MCHC)=29, serum iron=10ug/dl, ferritin=3.6 ng/dl].

Barium esophagogram was commented as normal. However endoscopy showed grayish thin arc-shaped membrane in the upper esophagus 20cm from the incisors (Figure 1), the scope did not pass further. Tests for other causes of anemia were negative.

Esophageal balloon dilation was done. After dilation (Figure 2), the scope was introduced easily and the rest of the esophagus, stomach and duodenum were all normal. The patient was given oral ferrous sulfate for six months until hematologic tests are normalized. The patient is doing well.

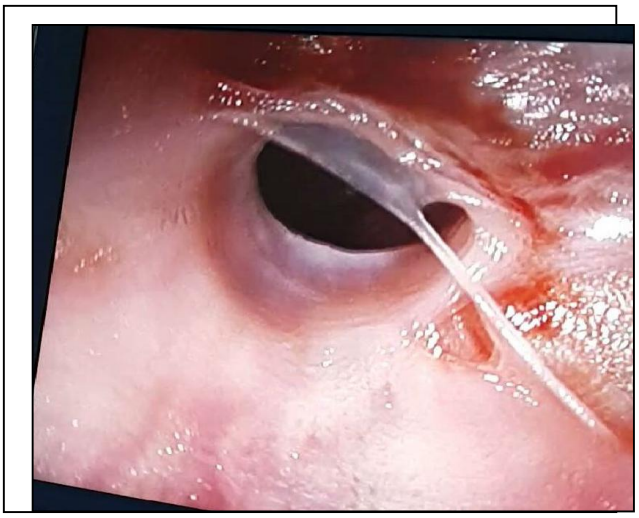

Figure 1: Upper esophageal web

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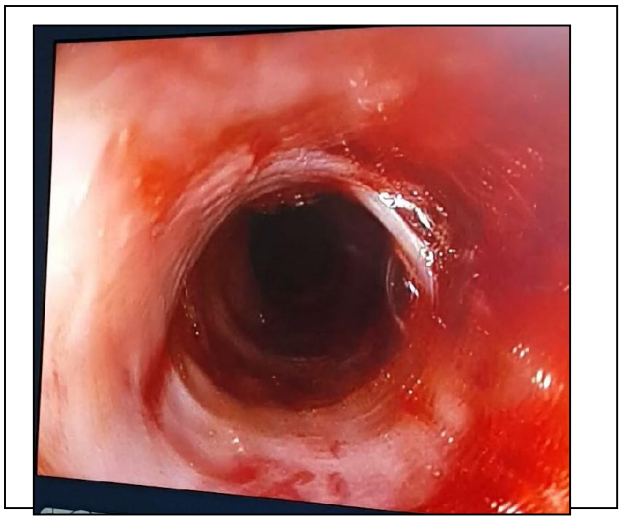

Figure 2: After endoscopic dilation

Case 2: A 25-year-old male presented with progressive difficulty of swallowing of solid and semi-solid foods for 15 years. He also had recurrent mouth ulcers, tinnitus, anorexia and weight loss. There was no history suggestive of choking episodes, ingestion of drugs or corrosives.

On physical examination, vital signs were normal. He had thin body built with pale conjunctivae. Oral examination showed glossitis and angular cheilitis (Figure 3). Examination of his extremities revealed koilonychias with pale fingernail beds.

Laboratory report showed iron deficiency anemia; Hb = 7.8 g/dl, MCV = 56.1fl, MCHC = 25.5 g/dl, serum iron = 11 ug/dl, ferritin=3.7 ng/dl and TIBC= 496 ug/dl. Barium esophagogram revealed post-cricoid short segment esophageal stricture. Upper gastrointestinal endoscopy showed an obstructing upper esophageal web (Figure 4). Biopsy was taken from the web and it showed a stratified squamous epithelium with normal range of maturation. This patient was treated with endoscopic bougie dilation and oral iron replacement. He is now in good health, after 2 years from treatment.

Figure3: Angular cheilitis

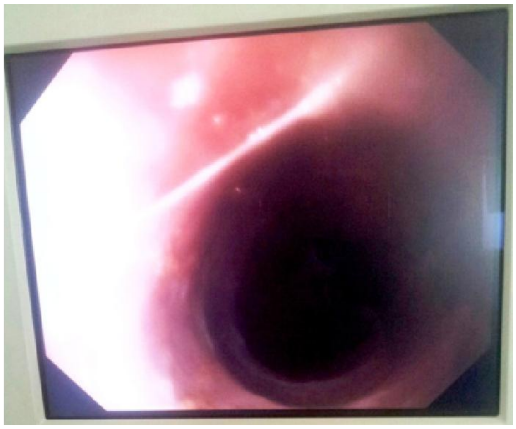

Figure 4: Esophageal web

Case 3: A 28-year-old lady from Addis Ababa came with difficulty of swallowing of one year duration. The dysphagia was limited to solid foods. She also experienced symptoms of anemia. She had no weight loss, overt gastrointestinal bleeding or abnormal menses. Physical examination showed pale conjunctivae. The vital signs were normal and the rest of the examination was unremarkable.

Laboratory investigations revealed hypochromic microcytic anemia: Hb=8.2 g/dl, MCV=68.5fl, MCHC=28.6 g/dl, serum iron=9 ug/dl, ferritin=4.88 ng/dl and TIBC = 337 ug/dl. Liver, kidney and thyroid function tests were all in the normal range. Stool exam for ova or parasite, fecal occult blood test and serology for HIV were negative.

Endoscopy showed upper esophageal arc-shaped webs (Figure 5). Through the scope balloon dilation was done and after esophageal dilation the rest of the esophagus, stomach and duodenum were normal. She was given oral ferrous agent for 6 months. After treatment, she has no dysphagia or symptoms of anemia and her laboratory tests are normal.

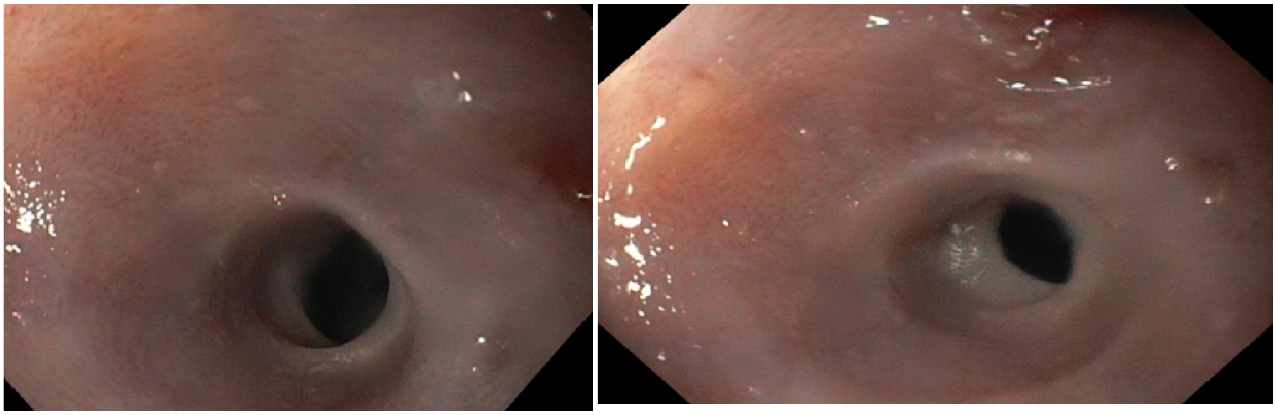

Figure 5: Upper esophageal webs

Case 4: A 39-year-old man came with difficulty of swallowing which has been there for one year. The dysphagia was only to solid meals. He had no anorexia or weight loss. He denied history of heartburn, regurgitation or corrosive ingestion. Examination showed mildly pale conjunctivae; otherwise, the rest of the examination was normal.

Laboratory findings revealed mild iron deficiency anemia: Hgb-10.2g/dl, MCV-72fL, and serum iron-17ug/dl. Organ function tests were in the normal range. Stool exam for parasites, fecal occult blood test and serology for HIV were negative.

On endoscopy, there were three semi-circumferential membranous webs in the upper esophagus about 20cm from the incisor teeth (Figure 6). Through the scope balloon dilation was done and the scope passed down easily. Biopsy showed stratified squamous epithelium with no dysplasia. The patient was given oral iron gluconate for 6 months. After treatment his symptoms completely resolved and anemia was corrected .

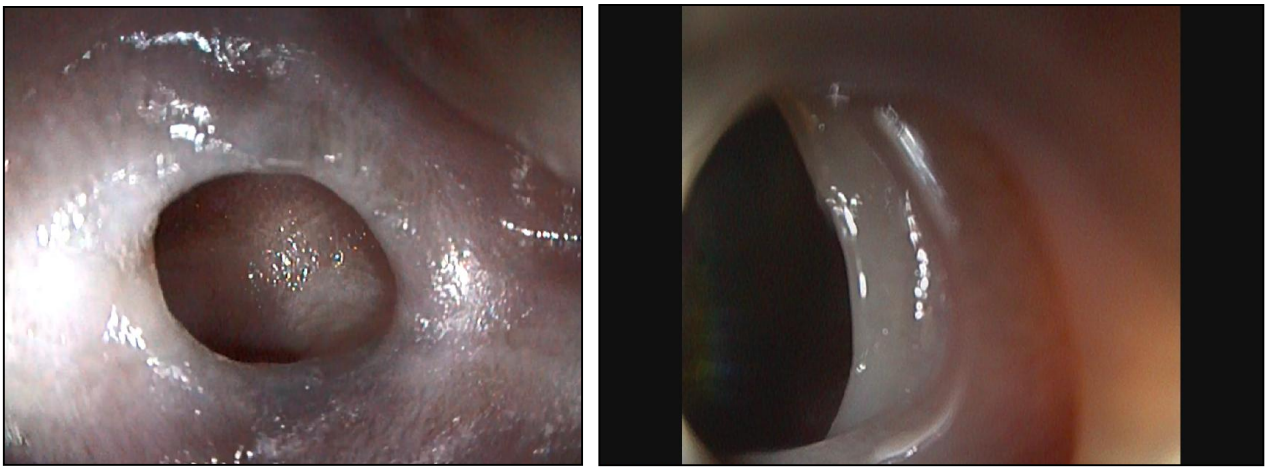

Figure 6: Upper esophageal webs

Table 1: Summary of the cases

	Case 1	Case 2	Case 3	Case 4
Age (years)	16	25	28	39
Sex	Female	Male	Female	Male
Duration of dysphagia	5 years	15 years	1 year	1 year
Hemoglobin (g/dl)	7.3	7.8	8.2	10.3
MCV (fL)	57.2	56.1	68.5	72
Iron (ug/dl)	10	11	9	17
Ferritin (ng/dl)	3.6	3.7	4.88	6.7

DISCUSSION

The classical triad of PVS includes upper esophageal web(s), iron deficiency anemia and dysphagia. Exact data about incidence and prevalence of the syndrome are not available. In the first half of the 20th century, Plummer-Vinson syndrome seemed to be common in Caucasians of Northern countries, particularly among middle-aged Scandinavian women (3). Currently, it is extremely rare due to better nutrition and health care condition in countries where the syndrome had been previously described (4). In a series of 1000 consecutive patients who underwent a cineradiographic examination of the hypopharynx and cervical esophagus, webs were found in 5.5% of the cases but only six patients had dysphagia attributable to the webs, and none of the patients fulfilled the criteria for Plummer-Vinson syndrome (5). In the recent years, instead of case series only case reports are published in the literature.

On our literature search of PVS in Africa, we found only a couple of reports. In a continent where both iron deficiency and malnutrition are common, the syndrome is thought to be very rare. However, it could be due to under-diagnosis or under-reporting.

Most of the patients with PVS are white middle-aged women, in the fourth to seventh decade of life (3,6) but the syndrome has also been described in children and adolescents (7-9). Analysis of English language case reports published in the literature from 1999 to 2005 revealed that 25 (89 %) out of the 28 adult patients with Plummer-Vinson syndrome were women (10). The mean age at presentation was 47 years (range 28–80 years). All patients had an iron deficiency anemia with a mean hemoglobin value of 8.2 g/dL.

The pathogenesis of PVS is not known but iron deficiency, auto immune processes and genetic predisposition have been proposed as possible etiopathogenetic factors. The alimentary tract is susceptible to iron deficiency; it rapidly loses iron dependent enzymes due to its high cell turnover, which then leads to mucosal degeneration and web formation. It was reported that in a patient with PVS, iron deficiency caused esophageal motility decrease; new motility studies showed normal amplitude of contraction after iron therapy (11).Plummer-Vinson syndrome may be accompanied by pernicious anemia, thyroiditis and celiac disease (12,13).

The most important symptom of this syndrome is dysphagia, which is usually limited to solid foods and is generally intermittent. Patients may also complain about choking and aspiration episodes (14). Symptoms and signs of iron deficiency anemia may also be present. Barium swallow studies and fluoroscopic evaluation suggest the diagnosis and the degree of stenosis. Esophago-gastro-duodenoscopy helps obtain histological samples to rule out other disorders, confirms the diagnosis and also helps therapeutically in the dilation of webs. Webs are tiny mucosal membranes covered by normal squamous epithelium.

The treatment of Plummer-Vinson syndrome is iron supplementation together with dilation therapy for patients who have profound dysphagia. Endoscopic dilation is simple and a chosen procedure in the treatment of the syndrome and upper esophageal web (15,16). Our patients were treated with oral iron replacement and endoscopic dilation. The anemia and dysphagia improved markedly after few weeks of therapy and none of the patients required repeat endoscopic dilation. This syndrome is known to be associated with upper alimentary tract cancer; esophageal or pharyngeal cancer can develop in 3% -15% of patients with PVS and surveillance endoscopy is recommended (17,18).

In conclusion, we would like to emphasize that Plummer -Vinson syndrome should still be considered in the differential diagnosis of dysphagia and it is important that it should be differentiated from other causes of dysphagia. Timely diagnosis and treatment of this syndrome is important as the treatment outcome is very good.

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