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Plummer-Vinson syndrome associated with lower esophageal ring (Schatzki ring) and hiatal hernia

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ABSTRACT

We report a case of Plummer-Vinson syndrome (PVS) and lower esophageal ring with a small sliding hiatal hernia. PVS is a rare entity formed by the combination of dysphagia, cervical esophageal web and iron deficiency anemia. It occurs mainly in middle-aged women[1,2,3]. A lower esophageal ring and a small sliding hiatal hernia were also observed in this case. We documented clinical manifestations of iron deficiency anemia through images and esophageal abnormalities through barium esophagogram.

Keywords: *Plummer-Vinson syndrome; Paterson-Brown-Kelly syndrome; sliding hiatal hernia; lower esophageal sphincter; iron deficiency anemia*

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CASE REPORT

A 39-year-old woman presented with dysphagia, odynophagia and weight loss. She reported that those symptoms started a long time ago, with gradual progression during the previous 6 months. She reported no heavy menstrual bleeding. Physical examination revealed nail deformity compatible with koilonychia (Figure 1a), angular cheilitis (Figure 1b) and atrophy of the tongue papillae (Figure 1c). Her body mass index (BMI) was 19.2 kg/m^2.

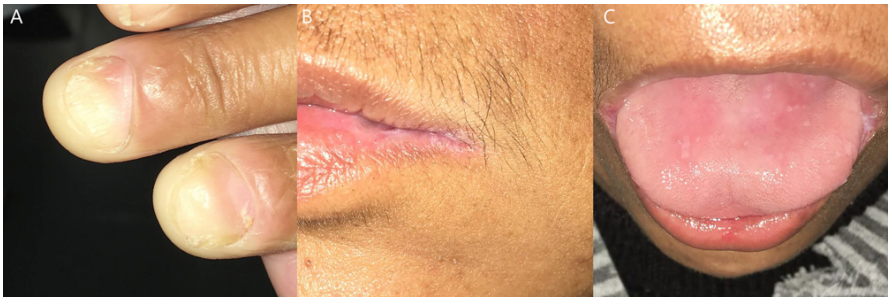

Figure 1: A: Koilonychia; B: Angular cheilitis; C: Atrophy of the tongue papillae.

Laboratory tests were performed, revealing microcytic hypochromic anemia and thrombocytosis. Hemoglobin level was 6.4 g/dL (reference range: 11.6-15.6 g/dL), hematocrit was 23.9% (reference range: 36.0-48.0%), mean corpuscular volume was 53 fL (reference range: 80-98 fL), red blood cell count was 4.52 million/µL (reference range: 4.0-5.4 million/µL), mean corpuscular hemoglobin was 14 pg (reference range: 28-32 pg), mean corpuscular hemoglobin concentration was 26.8 d/dL (reference range: 32-36 d/dL), red cell distribution width was 21.5% (up to 15.0%) and platelet count was 645.000/µL (reference range: 150.000-440.000/µL).

Other laboratory test results were as follows: serum iron: 6 µg/dL (reference range: 50-170 µg/dL); serum ferritin: 4 ng/mL (reference range: 10-291 ng/mL); transferrin saturation: 2% (reference range: 20-50%); total iron-binding capacity:

318 µg/dL (reference range: 250-450 µg/dL); vitamin B12: 260 pg/mL (reference range: 193-982 pg/mL); serum folate: 10.6 ng/mL (reference range: 3.0-17.0 ng/mL).

Because of the patient's obstructive symptoms and significant iron deficiency anemia, imaging studies of the upper gastrointestinal tract were performed. Digestive endoscopy was interrupted because of patient intolerance as well as resistance to the passage of the device in the cervical esophagus. Contrast radiography of hypopharynx, esophagus, stomach and duodenum showed membranes in the anterolateral and posterior walls, apparently forming a complete ring in the cervical esophagus, which determined a significant reduction in its lumen. There was also a lower esophageal ring approximately 5 mm thick and 5 mm in diameter and a small sliding hiatal hernia with the gastroesophageal junction, protruding 2.2 cm above the diaphragmatic hiatus (Figure 2).

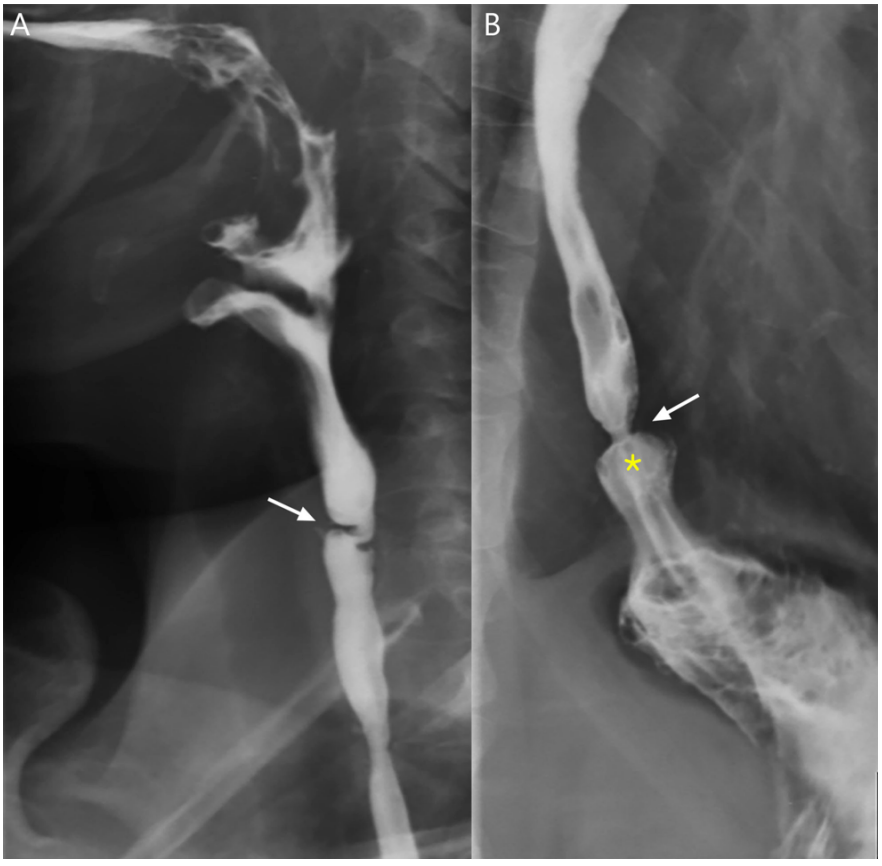

Figure 2: Barium esophagogram. A: Web *(arrow)* constricting the lumen of the esophagus; B: Prominent inferior esophageal ring *(arrow)* and small sliding hiatal hernia *(asterisks)*.

In the outpatient clinic, iron supplementation therapy was started and an upper endoscopy with esophageal dilation was ordered for further evaluation and treatment. Additionally, a medical appointment was scheduled for the patient after the procedure. Unfortunately, the patient did not attend the procedure nor the appointment. The follow-up was lost.

DISCUSSION

Plummer-Vinson syndrome (PVS), also known as Paterson-Brown-Kelly syndrome and sideropenic dysphagia[1], is a rare entity that occurs mainly in middle-aged women[1-3]. The disorder is characterized by the combination of dysphagia with esophageal web and iron deficiency anemia. An esophageal web is usually located in the proximal esophagus, consisting of a thin (1 mm to 2 mm) mucosal fold that typically appears as a diaphragm-like membrane. It is perpendicular to the long axis of the esophagus, constricting its lumen[4,6]. The esophageal web is usually semicircular and almost always arises from the anterolateral wall, but it may form a complete ring[1]. Patients with PVS have increased risk of developing carcinoma of the pharynx or esophagus[1,2].

Clinical manifestations of iron deficiency anemia are angular cheilitis, glossitis and koilonychia. Glossitis consists of painful tongue with papillae atrophy[3,5]. Koilonychia (spoon nails) is a less common

finding in physical examination but highly specific in iron-deficient states[6]. The patient presented with microcytic hypochromic anemia, reduced iron, ferritin and transferrin saturation and thrombocytosis. All of those laboratory abnormalities are manifestations of sideropenic anemia.

A Schatzki ring is a mucosal fold at the esophagogastric junction that may or may not be associated with symptoms[3,4]. It is usually symptomatic when the fold is thickened to 4 to 5 mm and the luminal aperture is narrowed to less than 12 mm. Clinical manifestations include intermittent episodic dysphagia to solids, often associated with substernal pain. The obstructive symptoms may be relieved by drinking liquids or regurgitating the ingested material. Occasionally, an impacted bolus must be removed through endoscopy[4].

The term lower esophageal ring was proposed in radiology because of a difficulty to distinguish a Schatzki ring from an annular peptic structure involving the esophagogastric junction[1]. Lower esophageal ring is associated with sliding hiatal hernia[1,4]. Gastric hernia occurs when the esophagogastric transition is above the esophageal hiatus. It affects about 10% of the population and prevalence increases with age[7]. In individuals in the 6th decade of life, the reported prevalence of these hernias is 50% to 60%[5,8]. Paul & Juhl report that small hiatal hernias are normal findings or changes associated with aging[9]. Radiographic criteria for the diagnosis of sliding hernia are gastroesophageal junction more than 2 cm above the hiatus[1,7], three or more gastric folds passing through the hiatus or diaphragmatic hiatus more than 3 cm in diameter[1].

Conflicts of Interest

The authors declare no conflicts of interest.

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