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Plummer-Vinson Syndrome and Role of Endoscopic Balloon Dilatation in a 4-Year-Old Child

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Abstract: Plummer-Vinson syndrome (PVS), also called Patterson-Kelly-Brown syndrome, is a rare cause of dysphagia in children. This syndrome is associated with single or multiple webs in the upper esophagus with frequent iron deficiency. PVS usually occurs in adults, particularly in Caucasian middle-aged women, in the fourth to seventh decade of life, and is rare in childhood. There are various theories about what causes PVS. One theory suggests that iron deficiency plays a crucial role in its development. Iron repletion often improves dysphagia, although some patients require esophageal dilatation or bougienage. Herein, we describe the case of a 4-year-old male child, having complaints of difficulty in swallowing solid food, diagnosed with PVS.

Key Words: Plummer-Vinson syndrome, dysphagia, anemia

INTRODUCTION

Plummer-Vinson syndrome (PVS), also called Patterson-Kelly-Brown syndrome, is a rare cause of dysphagia in children. This syndrome is associated with single or multiple webs in the upper esophagus with frequent iron deficiency (1). PVS usually occurs in adults, particularly in Caucasian middle-aged women, in the fourth to seventh decade of life (2); it is rare in childhood (3). Furthermore, the rapid decline in the prevalence of the syndrome is directly proportional to the improvement of nutritional status and the prevention of widespread iron deficiency in countries where the syndrome had been previously described (2). Herein, we describe the case of a 4-year-old male child, having complaints of difficulty in swallowing solid food, diagnosed with PVS.

Case

A 4-year-old boy presented with difficulty in swallowing solid food solid food, not associated with pain for the last 2 years. He also complained of a feeling of food impaction, followed immediately by vomiting. He was taking only liquids and semiliquid consistency food. Progressive pallor was also noticed by the father for the past 10 days associated with easy fatigability. There was no history of pica, bleeding manifestations, worm infestation, difficulty in breathing, or recurrent episodes of respiratory infections. On examination, the weight weight was 14.1 kg (between the 10th to 25th centile as per the World Health Organization growth chart) and height was 95 cm (less than the third centile as per World Health Organization growth charts). His heart rate was 152 beats per minute, his respiratory rate was 24 cycles per minute, his blood pressure was 102/74 mm Hg, and his oxygen saturation was 98% on room air. There was pallor and angular cheilitis but no edema or cardiomegaly. Systemic examination was normal. On investigations, hemoglobin was 3.9 g/dL (normal: 11–13 g/dL), mean corpuscular volume 53 fl (normal: 75–85 fl), mean corpuscular hemoglobin (MCH) 14.1 pg (normal: 24–30 pg), mean corpuscular hemoglobin concentration, 26.4 (normal: 32–36%) suggestive of microcytic hypochromic anemia, white cell count (WBC) 19,900 cells/mm^3 (polymorphs 43.2%, lymphocytes 49.4%, monocytes 5.2%) and platelet count was $1,308 \times 10^3$ cell/mm^3 (Normal: $150–450 \times 10^3$ cell/mm^3). Iron studies showed serum iron of 8 μg/dL (Normal: 33–193 μg/dL), total iron binding capacity of 653 μg/dL (Normal: 261–478 μg/dL), and transferrin saturation of 1.23% (Normal: 15–50%). The patient received 2 packed cell volume units before the upper gastrointestinal (GI) endoscopy. Direct laryngoscopy and barium swallow were normal (Fig. 1). Upper GI endoscopy showed an esophageal mucosal web 12 cm from incisors with difficulty in negotiating the 9.2 mm scope through the web (Fig. 2). Control radial expansion balloon catheter dilatation was done up to 10 mm. Postdilatation, the 9.2 mm scope could be easily negotiated through the web. The stomach as well as the first and second parts of the duodenum appeared normal except for pale mucosa. Although the narrowing persisted after the first session of endoscopy, the dysphagia decreased. The second session of control radial expansion balloon dilatation was performed after 3 weeks and was dilated up to 15 mm. There was no further complaint of dysphagia to solids postdilatation, adequate weight gain was noted during regular follow-ups and the patient was put on an iron-rich diet and iron supplements. Upper GI endoscopy has been advised after 1 year for cancer screening. The patient is currently being monitored on an outpatient basis.

DISCUSSION

Dysphagia in the upper cervical region is a rare symptom in children that requires explorations into its etiology. There are multiple causes that may lead to dysphagia, including benign strictures, esophageal, diverticula, esophageal peptic stenosis, congenital stenosis of the esophagus, eosinophilic esophagitis, hypertrophy of the cricopharyngeal muscle, achalasia, and PVS (1,2).

Among the various theories proposed to explain its development, one theory suggests that iron deficiency plays a pivotal role (4). Additionally, deficiencies in other essential vitamins, autoimmune factors, and potential genetic predisposition have also been considered as contributing factors (5). Iron insufficiency may cause a reduction in the activity of oxidative enzymes that rely on iron, which could lead to the development of webs due to muscle weakness and mucosal atrophy. This is supported by research that shows similar mitochondrial damage in the pharyngeal muscles of mice

Received August 2, 2023;accepted September 9, 2023.
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Authors declare no conflict of interest for this article. The article has been read and approved by all the authors, the requirements for authorship as stated earlier in this document have been met, and each author believes that the manuscript represents honest work.

JPGN Reports (2023) 4:4(e375)
ISSN: 2691-171X
DOI: 10.1097/PG9.0000000000000375

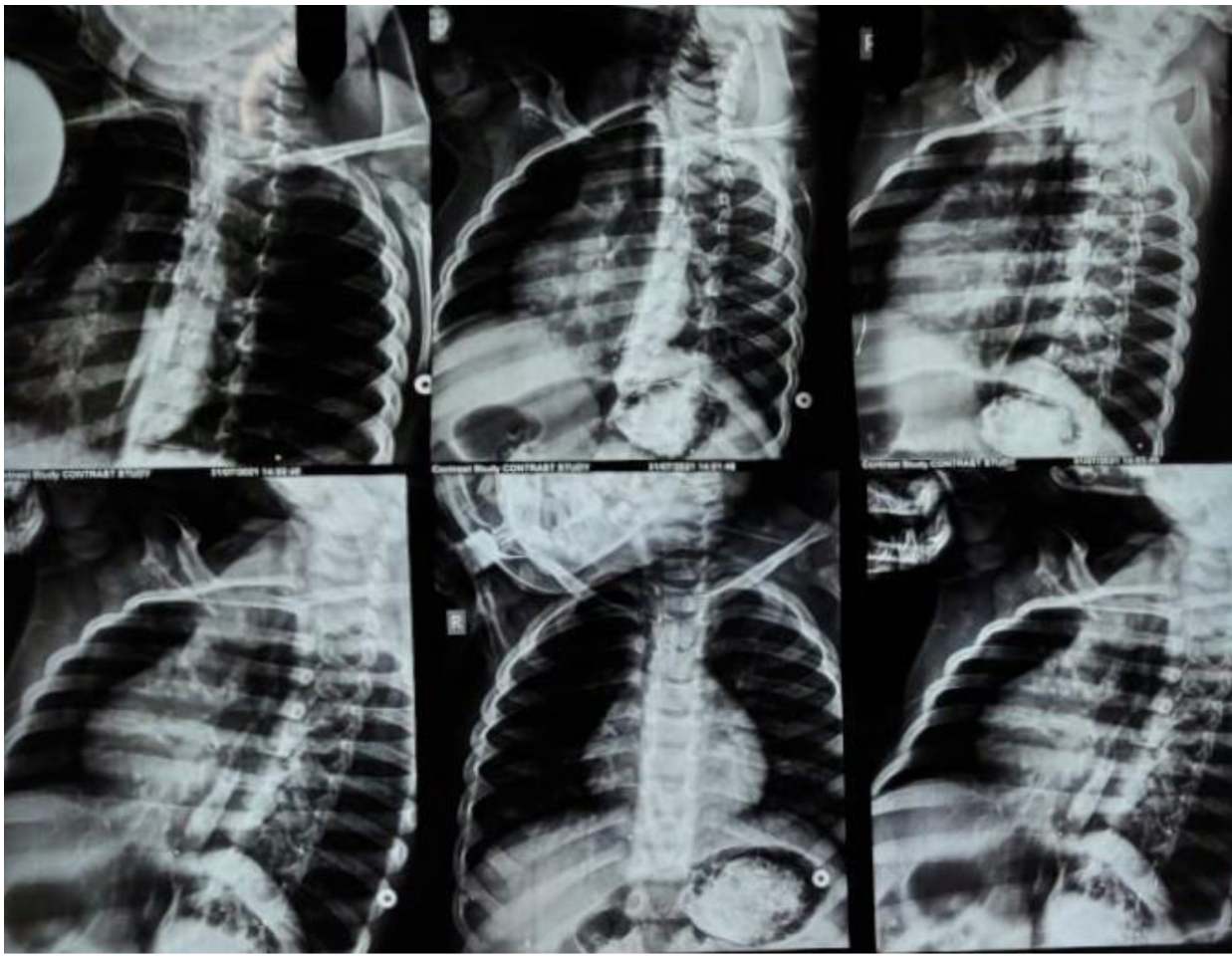

FIGURE 1. Barium swallow of the patient had no significant findings.

with iron-deficiency anemia, similar to progressive muscular dystrophy. These mitochondrial changes contribute to the degeneration of the nervous system and the aging of the mucosa (4).

PVS presents with progressive painless dysphagia, limited to solids, and associated with weight loss. The patient presents with features due to iron-deficiency anemia such as pallor, fatigue, weakness, and tachycardia. Additional features include angular cheilitis, koilonychia, and glossitis (2). Other physical findings include splenomegaly, and brittle nails (6). Our patient presented with dysphagia toward solids, fatigue, pallor, and features of angular cheilitis.

The diagnosis of PVS is based on the presence of iron-deficiency anemia and one or more postcricoid esophageal webs (6). Laboratory studies reveal iron-deficiency and microcytic hypochromic anemia. The webs may be visualized on barium swallow (1). Esophageal webs are thin, circular, or semicircular structures resembling a shelf, with a pinkish-white color and consists of 2 layers of mucosa with little fibrous tissue in between but without any muscular tissue. They are found only in the upper esophagus, typically situated just below the upper esophageal sphincter. Due to this location, obtaining a biopsy of a web can be challenging (7). The webs are, therefore, best diagnosed by cineradiography (6). In our patient, the barium swallow did not reveal any pathology.

The subsequent diagnostic test that should be considered is upper endoscopy or esophagoscopy. This procedure has the benefit of providing both diagnostic and treatment options simultaneously. The esophageal webs are extremely thin and situated near the upper esophageal sphincter, making them easy to overlook. Moreover, during the passage of the endoscope, there is a risk of accidentally rupturing the webs. Therefore, it is crucial to inform the endoscopist about the possibility of an esophageal web to improve the accuracy of this procedure (4), webs were revealed in the upper esophagus on endoscopy in our patient.

Iron repletion often improves dysphagia, although some patients require esophageal dilatation or bougienage. A multidisciplinary approach involving the primary care provider, gastroenterologist, hematologist, and radiologist can help provide holistic patient care with improvement in treatment and overall survival (4). Furthermore, the syndrome is associated with an increased incidence of postcricoid carcinoma, and surveillance endoscopy is recommended (6).

CONCLUSION

PVS is a complex disease process with only a handful of cases being reported in children. Current evidence points to good responses to the treatment of iron-deficiency and esophageal dilatation for dysphagia from esophageal webs, as demonstrated in our care. However, further studies are required to focus on the outcomes and prognosis of PVS in the pediatric population.

ACKNOWLEDGMENTS

Informed patient's guardian consent was obtained for publication of the case details.

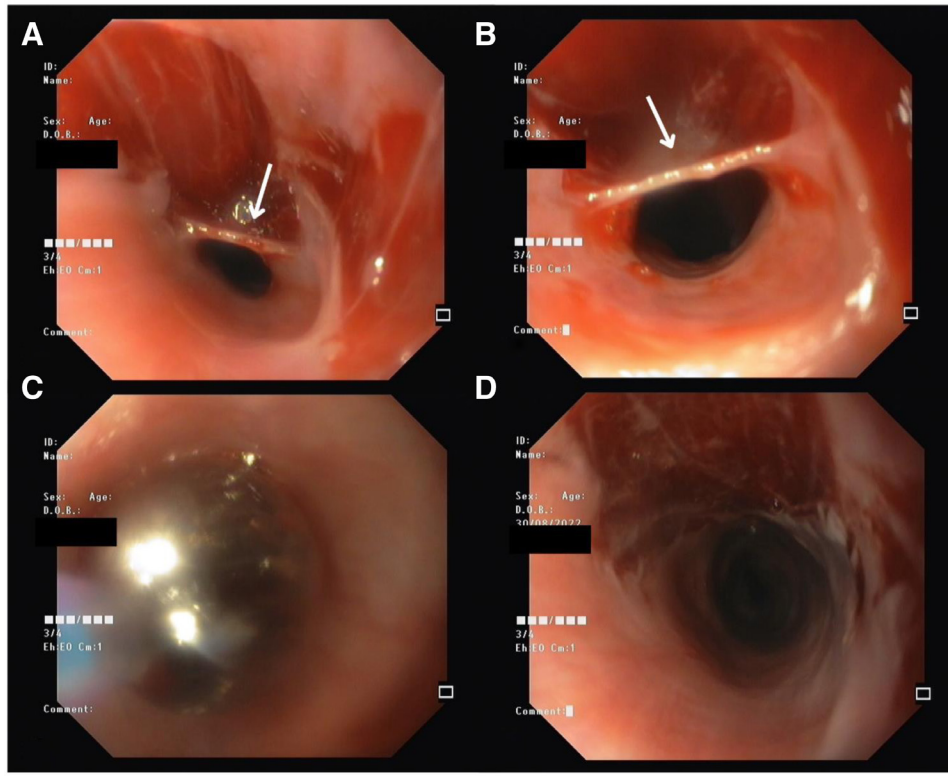

FIGURE 2. GI endoscopy showing esophageal webs (A,B), CRE balloon dilatation (C), postdilatation esophagus (D). CRE, control radial expansion; GI, upper gastrointestinal.

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