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Case Report

Plummer-Vinson Syndrome: A Rare Cause of Dysphagia in Children

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ABSTRACT

Plummer-Vinson Syndrome (PVS) or Paterson-Brown-Kelly syndrome is characterized by the classical triad of iron deficiency anemia, esophageal webs and dysphagia. PVS is commonly found in women of middle age especially in the fourth and fifth decade of life and is rarely reported in children. We report a case of a 4-year-old girl who had a classic presentation of PVS. Early diagnosis is of utmost importance for better prognosis as PVS is a precancerous condition with high malignant potential. Mainstay of management is correction of anemia by iron supplementation. Majority of the cases iron repletion improves the dysphagia. In some cases; esophageal dilatation is used to provide symptomatic relief.

Keywords: Dysphagia, Malignancy, Pediatrics, Iron deficiency anemia, Endoscopy.

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INTRODUCTION

Plummer-Vinson Syndrome (PVS) or Paterson-Brown-Kelly syndrome is a rare condition characterized by the classical triad of iron deficiency anemia, esophageal webs and progressive dysphagia.[1] Despite being first described as early as 1912, this condition still continues to remain enigmatic.[2] PVS is commonly reported in middle-aged Caucasian women (fourth to seventh decade of life).[1] It is important to diagnose PVS early as it is considered a risk factor for developing squamous cell carcinoma of the hypopharynx or upper esophagus.[3] Although rare, PVS has also been reported in children.[4] We herein report a case of PVS in a 4-year-old girl who presented with progressive dysphagia and was managed successfully.

CASE REPORT

A 4-year-old Asian girl presented with gradually increasing difficulty in swallowing solid foods and progressive weight loss for 6 months. No difficulty for swallowing liquid or semi-solid food was reported. There was no history of fever, cough, cold, throat pain or Koch's contact. As per the records, the child had lost 4 kg in the last six months prior to admission. On admission the heart rate was 104/min, respiratory rate was 22/min, blood pressure was 100/64 mmHg. Her weight was 10.2 kg (<5th centile for age and sex) and height was 92 cm. On examination, pallor was present. She also had angular cheilitis and koilonychia. Throat examination was normal. Rest of the general and systemic examination was normal. Investigations showed: hemoglobin 5.2 gm/dL, total leucocyte count 6200/cumm and platelet count 3.5 lac/cumm. Iron studies showed of low serum ferritin, high total serum iron binding capacity and low serum iron suggestive of severe Iron Deficiency Anemia (IDA). Thyroid function tests, celiac screen, chest X-ray, immunological tests and work-up for rheumatological conditions were normal. Barium swallow revealed a web in the hypopharynx at the level of C4-C5 vertebral bodies (Figure 1). Upper Gastrointestinal Endoscopy (UGE) showed a stricture just below the upper esophageal sphincter for which serial dilatations were done (Figure 2). Patient was started on iron supplements. Her dysphagia gradually improved and on follow up after 6 months, she remained asymptomatic. She had gained 5 kg weight on follow up.

DISCUSSION

Although the exact pathogenesis of PVS remains poorly understood, iron deficiency is considered as an important contributing factor.[1,2] Other etiologic factors include malnutrition, genetic predisposition [TMPRSS6 gene mutation], and autoimmune processes.[1-3] The main factor in all the above conditions is iron deficiency. Tissue iron plays an important role in the proliferation of epithelial cells. Due to high cell turnover, the epithelial layer of the upper alimentary tract is especially susceptible to iron deficiency.[5] Iron deficiency can lead to rapid loss of iron-dependent enzymes resulting in mucosal degenerations, atrophic changes and web formation. This can eventually lead to development of cancer of the upper

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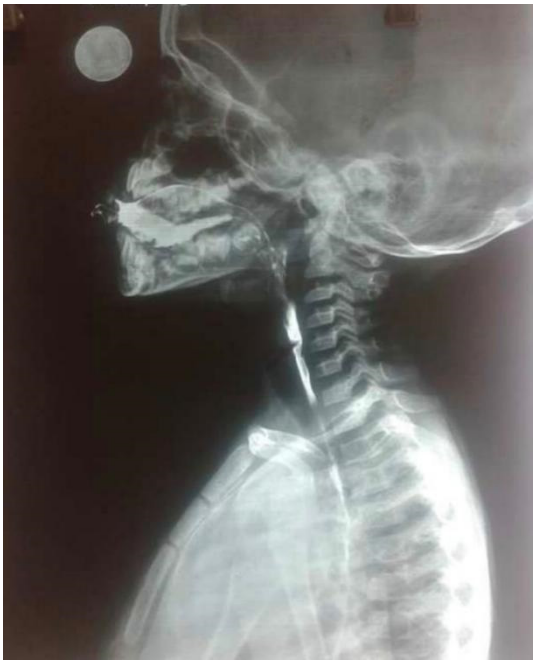

Figure 1: Barium swallow revealed a web in the hypopharynx at the level of C4-C5 vertebral bodies.

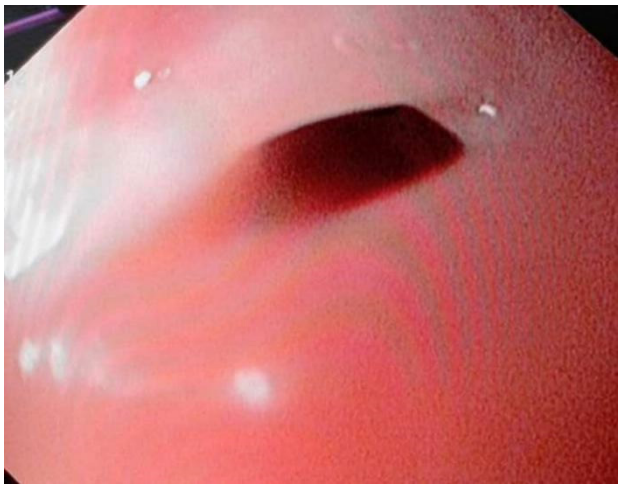

Figure 2: Upper Gastrointestinal Endoscopy (UGE) showed a stricture just below the upper esophageal sphincter.

gastrointestinal tract.[5] Around 3 to 15% of the patients, mostly women between 15 and 50 years of age, have been reported to develop esophageal or pharyngeal cancer.[1] The dysphagia in PVS is painless and intermittent or progressive over years, limited to solids in early phase and associated with weight loss. As the dysphagia is painless, patients usually tolerate it for long before presenting to the clinician. Patients may also have associated symptoms of iron-deficiency anemia like pallor, fatigue and weakness. Differentials include achalasia, reflux esophagitis, esophageal spasm, systemic sclerosis, Zenker's diverticulum. malignant tumors, benign strictures, diverticula, and neuromuscular disorders.[1-3]

Diagnosis of PVS is established by confirmation of iron deficiency anemia with demonstration of web by upper GI endoscopy. Management involves correction of anemia with iron supplements. In majority of the cases, dysphagia improves with iron supplements.[6] Iron therapy is advised even if the hemoglobin percentage is normal in the presence of web formation.[7] Cases in whom dysphagia does not respond to iron therapy will require endoscopic dilatation or incision.[1,6-8] We managed our case with both iron supplements and serial dilatation of the stricture. In view of the risk for malignant changes in upper gastrointestinal tract, patients will require regular follow-up. Currently, there are no formal guidelines for surveillance endoscopies to screen for squamous cell carcinoma. Some experts have recommended annual endoscopy for screening, while others recommend an endoscopic evaluation if symptoms return.[1,6-8]

CONCLUSION

Physicians should include PVS in the differential diagnosis of children presenting with dysphagia. Majority of the symptomatic patients require UGE with dilatation and iron replacement therapy. As patients with PVS are at increased risk of squamous cell carcinoma of the pharynx and the esophagus, regular follow up remains crucial.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

ABBREVIATIONS

PVS: Plummer-Vinson Syndrome; **IDA:** Iron Deficiency Anemia; **UGE:** Upper Gastrointestinal Endoscopy.

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