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Plummer-Vinson Syndrome

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(Plummer-Vinson Syndrome)

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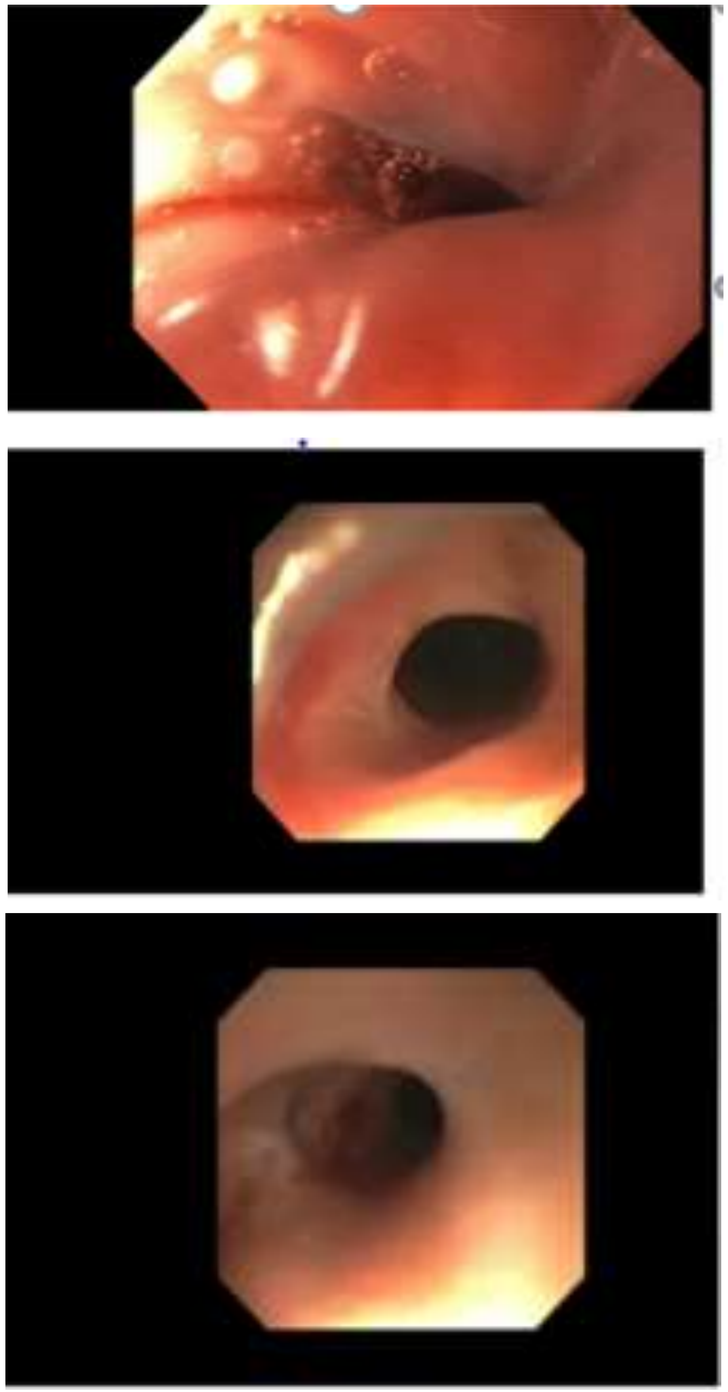
Internal Medicine

Background

- Plummer-Vinson syndrome (PVS) is a rare clinical disorder characterized by dysphagia, iron-deficiency anemia (IDA), and esophageal webs . It is typically a disease of middle-aged white women and usually presents with painless, progressive dysphagia limited to solids . Although PVS has been described from all parts of the world, the literature is still restricted to single case reports and small case series. While correcting the IDA has reproducibly been shown to alleviate dysphagia in most patients, the extreme rarity of the condition in individuals of African descent suggests a genetic component as well. We present the case of a young-aged African American woman with classic symptoms of PVS who had improvement in her dysphagia after dilation and iron replacement therapy and was found to have terminal ileum neuroendocrine tumor .

Case Presentation

37-year-old African American woman with no past medical history presented to the Emergency Department with a chief complaint of abdominal pain . She also reported associated fatigue and dysphagia. Over the past two months, she reported 40 Lbs. unintentional weight loss and decrease appetite. Her dysphagia was primarily to solids, which also had been worsening over the past few weeks. She had no history of similar symptoms in the past. She was not on any medications. On admission, vital signs were stable . In the Emergency Department,. Hemoglobin on admission was 3.3 g/dL [11.9–15.1 g/dL] with a mean corpuscular volume of 61.1 . Her serum iron on admission was 10 ug/dL [60–140 ug/dL], iron saturation was 2% [15–50%], and ferritin was 1.0 ng/mL [11.0–307 ng/mL]. Upon further questioning, the patient stated that she did not endorse menorrhagia. She denied hematemesis, hematochezia, or melena. She was transfused with 3 units of packed red blood cells and was started on 1000 mg of IV iron dextran complex infusions. Gastroenterology was consulted, and the patient underwent esophagogastroduodenoscopy (EGD) and colonoscopy. Colonoscopy showed erythematous and edematous mucosa in the terminal ileum which was Biopsied . The EGD revealed a few intrinsic stenoses in the upper esophagus which were successfully dilated with savary. A pathology exam of the terminal ileum showed well differentiated neuroendocrine tumor. Given the clinical, laboratory, and endoscopic (as shown below) Esophageal web located in the upper esophagus. a diagnosis of Plummer-Vinson syndrome was made. Her dysphagia improved over the three days in the hospital after iron infusions and blood transfusions, and she was able to tolerate a regular diet . Approximately 2 months after her discharge, a follow-up phone call was made to the patient. She stated that she had been compliant with all her medications and her dysphagia had resolved. At the time of the phone call, she was tolerating a regular diet without any dietary restrictions.



Upper third of esophagus on EGD

Discussion

Plummer-Vinson syndrome is a rare clinical disorder seldom seen in African American patients. The exact etiopathogenesis of how the triad of anemia, dysphagia, and esophageal webs develops remains speculative. However, it has been well established that PVS is most prevalent in white women ages 40–70 years old. In the African American population, which has relatively high rates of IDA, cases of PVS are fairly uncommon, making our case particularly unique. Additionally, there are several known comorbidities associated with PVS, including Celiac disease and menorrhagia, which may be underlying causes of IDA. However, to the best of our knowledge, there have been no reported cases of PVS with GI tumor. The causes of Plummer-Vinson syndrome and of web formation remain unknown. The most widely accepted theory of how PVS develops is that IDA plays a role in the development of dysphagia and esophageal web formation. It is hypothesized that IDA causes dysfunction of iron-dependent enzymes in the esophagus, which in turn causes oxidative stress and damages cellular DNA. This is thought to cause degeneration and ultimately mucosal web formation. The fact that dysphagia symptoms are often ameliorated by iron replacement therapy supports this theory. In reviewing the lecture multiple studies have shown the importance of Iron in the genome stability and the association of iron deficiency with increased GI malignancies. Moreover Preliminary evidence indicates that 20mg/day iron , may reduce the risk of GI cancer in the elderly as well as increasing genome stability in lymphocytes of children and adolescents. Damage to the genome has been linked to the origin and progression of many diseases and is the most fundamental pathology. Given the importance of iron for homeostasis and its potential influence over genome stability and cancer it is recommended to conduct further studies that conclusively define these relationships.

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