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Plummer-Vinson syndrome: is the immune system the missing piece?

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Abstract:

Plummer-Vinson syndrome (PVS) is a rare disorder characterized by a triad of iron-deficiency anemia, cervical dysphagia, and post-cricoid esophageal webs. The exact relationship between anemia and web formation remains unclear, with current hypotheses lacking strong evidence. Although treating anemia generally resolves dysphagia, some cases require endoscopic intervention. PVS is considered a precancerous condition due to its association with squamous cell carcinoma of the hypopharynx or upper esophagus. We present the case of a 53-year-old woman with psoriatic arthritis who exhibited iron-deficiency anemia unresponsive to oral iron and persistent dysphagia. Gastroscopy revealed esophageal narrowing, and she underwent successful balloon dilation following iron therapy. The patient is now asymptomatic. While the etiopathogenesis of PVS remains unknown, autoimmune conditions may play a role. Due to its potential link to neoplastic lesions, recognizing and managing PVS is critical. Further research is required to improve understanding and identify those at risk.

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Keywords: Plummer-Vinson syndrome. Anemia. Dysphagia. Iron-deficiency anemia. Paterson-Kelly syndrome. Immune system.

Dear Editor,

Plummer-Vinson syndrome (PVS) is a rare condition characterized by the triad of iron-deficiency anemia, cervical dysphagia, and post-cricoid esophageal webs. Although the association between anemia and web formation is recognized, the underlying mechanism remains poorly understood. While PVS generally has a favorable prognosis, as treating the anemia often resolves dysphagia, some cases may require endoscopic dilation. Additionally, PVS is considered a precancerous condition due to its association with an increased risk of squamous cell carcinoma of the hypopharynx or upper esophagus, although there is no established consensus on surveillance and follow-up strategies.

Case Report

We present the case of a 53-year-old woman with a history of psoriatic arthritis treated with methotrexate, etoricoxib, prednisone, and folic acid. She was referred for evaluation of symptomatic iron-deficiency anemia unresponsive to oral iron, along with oropharyngeal dysphagia for several months. The absence of overt bleeding prompted further investigation with CT enterography and colonoscopy, both of which revealed no significant findings. Gastroscopy revealed a fibrous ring causing narrowing of the esophageal lumen 15 cm from the dental arch, necessitating the use of a pediatric endoscope to complete the examination.

Based on these findings and the suspected diagnosis of PVS, intravenous iron therapy was initiated, followed by oral supplementation. This led to normalization of laboratory values but persistent dysphagia. Therapeutic endoscopy with 14 mm

balloon dilation was performed. A follow-up barium swallow study demonstrated good passage of contrast with no evidence of esophageal defects or stenosis.

The patient remains under close follow-up every six months, is currently asymptomatic, and no longer requires iron supplementation. Given the favorable clinical course, a surveillance endoscopy is planned in three years.

Discussion

The etiopathogenesis of PVS, first described over a century ago, remains unclear. Despite its rarity, even in regions with high iron-deficiency anemia prevalence, autoimmune factors may contribute to its development. Conditions like systemic sclerosis and eosinophilic esophagitis share chronic inflammation, with elevated proinflammatory cytokines (TNF-α, IL-6, IL-1β) and autoantibodies, leading to esophageal fibrosis and tissue remodeling. In PVS, chronic inflammation can be exacerbated by iron deficiency, which plays a key role in immune dysfunction by increasing cytokine production and perpetuating systemic inflammation.

In the presented case, the patient was on methotrexate and prednisone, immunomodulators that may have prevented PVS relapse. This syndrome's association with neoplastic lesions highlights the need for further investigation into its pathophysiology, particularly regarding the interaction between iron deficiency, inflammation, and autoimmune disorders.

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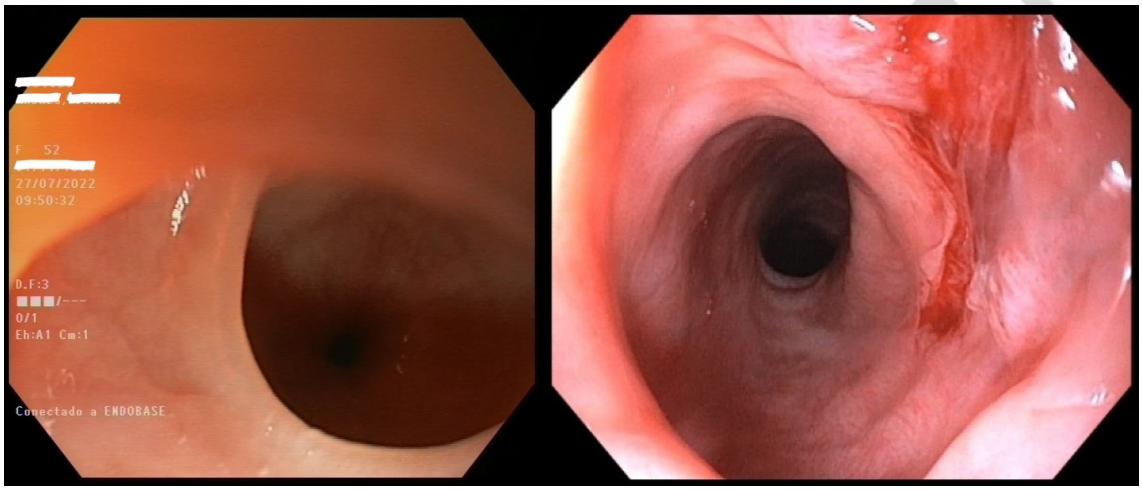

Figure 1: Left: cervical esophageal web. Right: Esophagus after endoscopic dilatation.