EDITORIAL

Plummer-Vinson Syndrome

Plummer-Vinson Syndrome can be associated with celiac disease, chronic disease, Thyroid disease, Rheumatic arthritis. Plummer-Vinson Syndrome (PVS) is a rare condition characterized by the classic triad of dysphagia, iron-deficiency anemia, and esophageal webbing. Plummer-Vinson Syndrome is more common in middle-aged women and is associated with an increased risk of developing squamous cell carcinoma of the pharynx and proximal esophagus.

Introduction:
Plummer-Vinson syndrome (PVS) is a rare condition characterized by the classic triad of post-cricoid dysphagia, iron-deficiency anemia, and upper esophageal webs.[1] In the United Kingdom, it is known as Paterson-Brown-Kelly syndrome. This name was given after two British laryngologists, Donald Ross Paterson (1863-1939) and Adam Brown-Kelly (1865-1941), who published their findings in 1919. PVS is more common in middle-aged women. The etiology of Plummer-Vinson syndrome remains unknown. Proposed etiopathogenic mechanisms include nutritional and iron deficiencies, genetic predisposition, and autoimmunity.[2] PVS was commoner in the earlier half of the 20th century, particularly in middle-aged Scandinavian women. In earlier Scandinavian studies, up to 90% of patients with PVS were reported to be women. The typical age at diagnosis is between 40 and 70 years. A limited number of cases are seen in children.

The pathogenesis of Plummer-Vinson syndrome and the formation of the esophageal web is not well known. It has been postulated that iron deficiency induces iron-dependent enzyme dysfunction, leading to oxidative stress and DNA damage.[4] The malfunctioning of iron-dependent oxidative enzymes produces myasthenic changes in muscles responsible for swallowing, atrophy of the esophageal mucosa, and formation of webs as epithelial complications.[5] Repeated injury to epithelia due to iron deficiency leads to atrophy of mucosa and degradation of pharyngeal muscles, leading to the development of esophageal webs. The esophageal web is localized below the cricopharyngeal muscle and is asymmetrically attached to the anterior esophageal wall. The esophageal web is a thin mucous membrane composed of squamous epithelia.[4]

The role of mucosal inflammation and atrophy, especially in the post-cricoid region, has been suggested as a factor for the pathogenesis of PVS. The post-cricoid region experiences maximum trauma while swallowing the solid bolus, leading to an increased risk of web formation.

PVS has also been proposed as an autoimmune phenomenon. It has been associated with autoimmune pathologies, such as pernicious anemia, rheumatoid arthritis, celiac disease, and thyroiditis.

Long-standing iron deficiency anemia can present as dyspnea, tachycardia, weakness, pallor, and koilonychia. Dysphagia is painless and slowly evolving, starting with solid foods and difficulty swallowing liquids after years of initial onset.

Although Plummer-Vinson syndrome is a rare diagnosis, there should be high clinical suspicion in patients with iron-deficiency anemia, esophageal webs, and post-cricoid dysphagia. Hematological testing is done to ascertain the cause of iron deficiency and the severity of anemia. Complete blood count, peripheral smear, and iron studies (e.g., serum iron, ferritin, total iron-binding capacity [TIBC], transferrin satu-

ration) should be carried out to confirm the diagnosis of iron deficiency anemia.

The esophageal web is investigated by radiographic tools such as barium swallow, which is generally available easily even in remote locations and is a diagnostic tool with the advantage of reproducible documentation. Videofluoroscopy is usually more reliable for the demonstration of esophageal webs.[6]

Medical management of Plummer-Vinson syndrome includes iron supplementation. Occult or overt blood loss is generally ruled out, along with any underlying malignancies or iron malabsorption. Iron replacement is essential to correct anemia and to resolve most of the features associated with iron deficiency. Dysphagia in many patients resolves with just iron supplementation.[8] However, dysphagia caused by more advanced disease is unlikely to respond to medical management alone and, thus, is managed with endoscopic dilation. it will require special treatment, such as celiac disease. Aside from iron replacement, dietary modification is sufficient in mildly symptomatic patients. Those with advanced and long-standing dysphagia typically require mechanical dilation. Commonly used techniques include endoscopic balloon dilatation or Savary-Gilliard dilators.[9] In a prospective study, Goel et al. observed that esophageal-web-related dysphagia in PVS responded well after one session of endoscopic dilation.

Prognosis:
Patients with Plummer-Vinson syndrome have an excellent outcome, with most symptomatic patients requiring only one esophagogastroduodenoscopy with dilatation for complete relief of symptoms in conjunction with iron replacement therapy.

Patients are at an increased risk of developing squamous cell carcinoma of the hypopharynx or upper esophagus, which may be related to chronic iron deficiency. This is believed to cause irreversible mucosal changes leading to malignant degeneration.[17]

Complications:
Untreated esophageal webs can lead to dysphagia for solids, absolute dysphagia, and aspiration pneumonia.

Iron-deficient patients may develop symptomatic anemia (fatigue, malaise, dyspnea, angina pectoris) if iron supplementation is not provided.

Patients may develop squamous cell cancer of the proximal esophagus, although the actual risk is unknown.

Endoscopic treatment of the esophageal web with Savary dilatations or balloon dilatations can be associated with a small risk of esophageal perforation.

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