Indian Journal of Nursing Sciences

International Peer Reviewed Journal

Mini Review Article

Plummer-Vinson Syndrome – A Community-Based Case Study

Joy Priyadharishini, Liza L

Department of Community Health Nursing, College of Nursing, Christian Medical College, Vellore, Tamil Nadu, India

Abstract

Plummer-Vinson syndrome (PVS) is a rare syndrome mostly affecting women, which is mainly due to dietary deficiencies. Health education and health promotion are the key in prevention of this syndrome. Systematic history collection and physical examination will help the health-care provider to detect the syndrome and start treatment at the earliest. Negligence of the disease or mismanagement might lead to devastating complications. The condition has been reported most commonly in thin-built, middle-aged, and American women. The esophageal webs in PVS are thin mucosal folds, which are best seen either in lateral views at barium swallow or at esophagoscopy. These are usually semilunar or crescentic, being located most often along the anterior esophageal wall, but can be concentric. The exact cause and pathogenesis of PVS remain unclear, though iron and other nutritional deficiencies, genetic predisposition, and autoimmunity have all been implicated in formation of the webs. Treatment includes correction of iron deficiency and endoscopic dilation of the esophageal webs to relieve dysphagia. PVS is associated with an increased risk of hypopharyngeal and esophageal malignancies. Correction of iron deficiency may arrest and reverse the mucosal changes and possibly reduces this risk. This article discusses on the etiology, clinical manifestations, investigations, management, and complications of PVS. Primary health-care professionals have a greater role in primary prevention and early detection of this syndrome thereby manages the symptoms well.

Key words: Dysphagia, esophageal web, iron-deficiency anemia, postcricoid dysphagia

Address for correspondence: Joy Priyadharishini, Department of Community Health Nursing, College of Nursing, Christian Medical College, Vellore, Tamil Nadu, India. E-mail: joysherwyn.js@gmail.com

Introduction

Plummer-Vinson Syndrome (PVS) presents as a classical triad of dysphagia, iron-deficiency anemia (IDA), and esophageal web. It is a rare medical condition and mostly affecting women in the age group of 40–70. The affected individuals are at an increased risk of Squamous cell carcinoma of the pharynx and the esophagus. The syndrome is named after two physicians, Henry Stanley Plummer (1874–1936) and Porter Paisley Vinson (1890–1959). The early case reports by Dr. Plummer and Dr. Vinson were patients with long-standing iron deficiency, dysphagia, spasm of the upper esophagus without anatomic stenosis, and "angulation" of the esophagus. It is also termed as "Paterson-Brown-Kelly syndrome" after two British laryngologists; Dr. Donald Ross Paterson and Dr. Adam Brown-Kelly, who published similar findings in the year 1919 independently. Dr. Paterson was the first to suggest the association with post-cricoid carcinoma.[1] PVS is a rare condition that continues to be enigmatic, even though a century has passed since its first description.[2] The literature available on its pathogenesis, treatment and natural history is limited to case reports, retrospective short case series and a few large series.[3,4] In this review, we have attempted to collate the available information on PVS.

Access this article online	
Website:http://www.innovationalpublishers.com/Journal/ijns	e-ISSN: 2581-463X
DOI: 10.31690/ijns.2021.v06i01.006	

How to cite this article: Priyadharishini J, Liza L. Plummer-Vinson Syndrome–A Community Based Case Study. Indian Journal of Nursing Sciences 2021;6(1):30-33.

Definition

PVS is defined by the classic triad of dysphagia, IDA and esophageal webs. Even though the syndrome is very rare

nowadays, its recognition is important because it identifies a group of patients at increased risk of squamous cell carcinoma of the pharynx and the esophagus.[5]

Etiology

PVS is a rare condition, and the data on its incidence or prevalence rate are limited to a single population-based study conducted in South Wales in early 1960s. In this study, 1994 men and 2346 women were screened for post-cricoid dysphagia using a questionnaire followed by a barium study in those reporting symptoms. The prevalence of post-cricoid webs was found to be 0.3–1.1% and 8.4–22.4% among women overall and in women with dysphagia, respectively. None of the men were found to have a post-cricoid web.[6] The proposed causative mechanisms have included IDA, malnutrition, genetic predisposition, autoimmune processes, and other dietary deficiencies (such as of pyridoxine or riboflavin).[6]

Pathogenesis

The pathogenesis of PVS is unknown. The most important possible etiological factor is iron deficiency. This theory is primarily based on the finding that iron deficiency is a part of the classic triad of PVS together with dysphagia and esophageal webs and that dysphagia can be improved by iron supplementation. Indeed, impaired esophageal motility has been described in PVS and it was corrected by iron treatment. It has been shown that iron deficiency can precede dysphagia.[5]

On the other hand, the alimentary tract is susceptible to iron deficiency; it rapidly loses iron-dependent enzymes due to its high cell turnover, which is speculated to cause mucosa degeneration and web formation. However, large clinical series suggested that for many patients iron deficiency is neither a necessary nor a sufficient cause of web formation. Other etiologic factors include malnutrition, genetic predisposition or even autoimmune processes such as celiac disease, thyroid disease, and rheumatoid arthritis.[5]

Clinical Description

PVS has been identified as a risk factor for developing squamous cell carcinoma of the upper gastrointestinal tract. 3–15% of the patients with PVS, mostly women between 15 and 50 years of age, have been reported to develop esophageal or pharyngeal cancer. A decreasing trend in the overall incidence of hypopharyngeal cancer in women was demonstrated, probably due to diminished prevalence of PVS.

The main clinical features of PVS are postcricoid dysphagia, upper esophageal webs and IDA. The dysphagia is usually painless and intermittent or progressive over years, limited to solids and sometimes associated with weight loss. Symptoms resulting from anemia such as weakness, pallor, fatigue, and tachycardia may dominate the clinical picture. Furthermore, it is characterized by glossitis, angular Cheilitis and Koilonychia (spoon-shaped finger nails). Enlargement of the spleen and thyroid may also be observed.[5]

Investigations

The objectives of investigations in a patient with suspected esophageal web or PVS are to:

- Diagnose anemia
- Ascertain the cause of anemia
- Assess the severity and cause of dysphagia
- Localize the obstructing lesion.

Confirmatory tests

- Hematologic tests
- Radiographic (Barium Swallow).

Hematologic tests typically reveal IDA with decreased values of hemoglobin, hematocrit, mean corpuscular volume, serum iron and ferritin, and increased total iron binding capacity. Further laboratory abnormalities are usually not described.

Barium swallow radiography is the investigation most commonly asked for if an esophageal web is suspected. It has a few advantages over endoscopic examination. Besides being time-honored, it is more easily available in remote locations and can be interpreted by a radiologist or a clinician with no special skill or training. Further, it helps to differentiate between benign and malignant causes of obstruction, planning of definitive treatment, and provides a reproducible documentation of pretreatment status for comparison after treatment or other later use.

Endoscopic examinations of the esophagus may be required to identify associated conditions such as thyroid disorders, celiac disease so on to exclude hypopharyngeal or esophageal malignancy, and to identify the cause.[1]

Diagnosis

The diagnosis is based on the evidence of IDA and one or more esophageal webs in a patient with postcricoid dysphagia. Esophageal webs can be detected by barium swallow X-ray but the best way for demonstration is the videofluoroscopy. Webs are also detectable by upper gastrointestinal endoscopy. They appear smooth, thin, and gray with eccentric or central lumen. The webs typically occur in the proximal part of the esophagus and may be missed and accidentally ruptured unless the endoscope is introduced under direct visualization.[7]

The esophageal webs, which can also occur in the absence of anemia and PVS, are characterized by one or more thin horizontal membranes consisting of squamous epithelium and submucosa. They usually protrude from the anterior

wall, extending laterally but not to the posterior wall, which means that they rarely encircle the lumen.

Differential Diagnosis

Since dysphagia is a main clinical feature of PVS, the differential diagnosis includes all other causes of dysphagia especially malignant tumors, benign strictures, or esophageal rings. Other reasons for dysphagia are diverticula, motility disorders such as achalasia, spastic motility disorders, scleroderma, diabetes mellitus, gastroesophageal reflux disease, and neuromuscular and skeletal muscle disorders.[1]

Histopathology

Histologically, webs in patients with PVS show fibrosis, epithelial atrophy, epithelial hyperplasia and hyperkeratosis, basal cell hyperplasia, and some features of chronic inflammation.[1]

Treatment

Medical management

- Taking iron supplements may improve the swallowing problems
- If supplements do not help, the web of tissue can be widened during upper endoscopy. This will allow you to swallow food normally
- Instructing the patient to chew food thoroughly is usually the only treatment required in wider rings, but narrow-lumen rings require dilation by endoscopy or bougienage
- Surgical resection is rarely require.[1]
    - Devices used to stretch the esophagus (dilators) may cause a tear. This can lead to bleeding. PVS has been linked to esophageal cancer.

Prognosis

Patients with PVS have an excellent outcome. Patients are at an increased risk of developing squamous cell carcinoma of hypopharynx or upper esophagus. Long-standing iron deficiency is assumed to cause an irreversible mucosal change which potentially leads to malignant degeneration.[1]

Complications

If untreated it carries a high risk of progression to squamous cell cancer of the esophagus. Esophageal webs if untreated may also cause significant obstruction of the lumen and persistent dysphagia.[1]

Case History

Mrs. Vasanthi, a 33-year-old lady presented with complaints of slowly progressing dysphagia (difficulty in swallowing solid food) and weight loss of 3 kg in 2 months. History collection revealed presence of hypothyroidism, chronic anemia, and short menstrual cycle. Laboratory hematologic tests showed IDA. The patient first visited a physician for evaluation of dysphagia. A laryngoscopy was performed to rule out any obstruction or growth in the throat, which revealed no abnormalities. The patient was further referred to a gastroenterologist for evaluation of dysphagia, since she was suspected for the presence of esophageal web. The investigations including radiographic (barium swallow) and endoscopic examinations of the esophagus were done. The esophageal web was confirmed through barium X-ray and it was successfully disrupted without any complications. She was advised to take iron supplements and review gastroenterologist once in 2 years to avoid complications and facilitate early diagnosis of esophageal cancer. In this case, the patient had a recurrent esophageal web which was growing every 2 years and required a gastrointestinal endoscopy to disrupt the web. This history reveals that a patient with PVS requires periodic follow-up and evaluation of symptoms.

Role of a Community Health Care Provider

A primary health-care provider has the prime responsibility to deliver holistic care to the people at the community level. It is essential to create awareness to the public and involve every health-care stakeholder in making policies and recommendations to combat the problem. Some of the responsibilities of the primary health-care provider are:

- Malnutrition must be prevented or managed at the earliest to prevent PVS
- Providing health awareness on intake of nutritional iron supplements
- As deficiency of pyridoxine or riboflavin is a predisposing factor for PVS, any nutritional deficiencies must be detected at the earliest and handled
- Improvise the existing national nutritional health programs to benefit the people in need
- When genetic predisposition is detected; care should be taken so that the couples are counseled adequately
- Prompt referral services for cases which could not be handled at the primary level.

Conclusion

PVS is a rare uncommon disease. The commonness of IDA among adult women and the impact of cancer on an individual's life make it necessary to create awareness about the disease. The symptoms are definite but many a times it is left unnoticed. In a way, early diagnosis and treatment avoids recurrence of the disease. The treatment and prevention completely depends on the individual's lifestyle modifications including nutritious diet, observing for dysphagia and fatigue.

Acknowledgment

We would like to acknowledge all the community health nurses who provide tireless services to the people in the primary health-care setting.

References

1. Verma S, Mukherjee S. Plummer Vinson Syndrome. Treasure Island, FL: Stat Pearls; 2019.
2. Plummer HS. Diffuse dilatation of the esophagus without anatomic stenosis (cardiospasm): A report of ninety-one cases. J Am Med Assoc 1912;58:2013-5.
3. Goel A, Lakshmi CP, Bakshi SS, Soni N, Koshy S. Single-center prospective study of Plummer-Vinson syndrome. Dis Esophagus 2016;29:837-41.
4. Bakari G, Benelbarhdadi I, Bahije L, El Feydi Essaid A. Endoscopic treatment of 135 cases of Plummer-Vinson web: A pilot experience. Gastrointest Endosc 2014;80:738-41.
5. Novacek G. Plummer-Vinson syndrome. Orphanet J Rare Dis 2006;1:36.
6. Goel A, Bakshi SS, Soni N, Chhavi N. Iron deficiency anemia and Plummer-Vinson syndrome: Current insights. J Blood Med 2017;8:175-84.
7. Lynch KL. Lower Esophageal Ring; 2019. Available from: https://www.msdmanuals.com/home/digestive-disorders/esophageal-and-swallowing-disorders/lower-esophageal-ring. [Last accessed on 2019 Dec 20].