Case Report

Indian Journal of Dental Education
Volume 12 Number 4, October - December 2019
DOI: https://dx.doi.org/10.21088/ijde.0974.6099.12419.7

Plummer–Vinson Syndrome: A Rare Entity–Triad with Malignant Potential

Asish Rajasekharan[1], Sherin Ann Thomas[2], Anita Balan[3], Vinimol Chandini[4], Twinkle S Prasad[5]

Author's Affiliation: [1]Associate Professor and Head, [2]Assistant Professor, Department of Oral Medicine & Radiology, Government Dental College, Alappuzha, Vandanam, Kerala 688005, India. [3]Principal, Government Dental College, Chalakkuzhi, Thiruvananthapuram, Kerala 695011, India. [4]Assistant Professor, Department of Pharmacology, Government Medical College, Kollam, Kerala 691574, India. [5]Associate Professor, Dept. of Oral Medicine & Radiology, Government Dental College, Gandhinagar, Kottayam, Kerala 686008, India.

Corresponding Author: Asish Rajasekharan, Associate Professor and Head, Department of Oral Medicine & Radiology, Government Dental College, Alappuzha, Vandanam, Kerala 688005, India.

E-mail: asishrajasekharan12@gmail.com

Received on 24.10.2019; **Accepted on** 27.11.2019

How to cite this article:
Asish Rajasekharan, Sherin Ann Thomas, Anita Balan, et al. Plummer–Vinson Syndrome: A Rare Entity–Triad with Malignant Potential. Indian J Dent Educ. 2019;12(4):157-160.

Abstract

Plummer–Vinson syndrome is a disease characterized by iron-deficiency anemia, difficulty in swallowing, glossitis, cheilosis and esophageal webs.[1] Treatment with iron supplementation and mechanical widening of the esophagus generally provides an excellent outcome.

Keywords: Plummer–Vinson syndrome; Iron-deficiency anemia; Esophageal web; Glossitis.

Introduction

The relative incidence of this syndrome has become rare. It is also termed as Sideropenic Dysphagia, Paterson–Brown–Kelly syndrome.[1–3] The reduction in the prevalence of Plummer–Vinson syndrome has been hypothesized to be as the result of improvements in nutritional status.[1–3] This syndrome generally occurs in postmenopausal women.[2–5.] Its identification and follow-up is considered relevant due to increased risk of squamous cell carcinomas of the esophagus and pharynx.[4–10] The condition is associated with koilonychia, lemon tinted pallor of skin, glossitis, cheilitis, and splenomegaly.[4,5,11–15] Esophageal webs in Plummer–Vinson syndrome are found at upper end of esophagus post-cricoid region may be found at the lower end of esophagus.[8–10]

Case Report

The case report is based upon a 65-year-old female patient who was referred from the ENT department for routine dental check up before surgical treatment planning. The complaint started two years back with difficulty in swallowing food. Patient consulted ENT surgeon for the problem of hoarseness of voice and pain on swallowing food. Patient had consulted the dermatologist for the evaluation of asymptomatic nail change.

The patient didn't have any ill habits. She had a non-vegetarian diet and had apparently normal sleep, bladder and bowel habits. She had reduced appetite since 18 months. The rest of the history is non-contributory.

General examination revealed apparently built and nourished adult female with normal gait and posture. There were symptoms of anemia and lemon tinted pallor of facial skin (Fig. 1). The examination of gastrointestinal system revealed dysphagia resulting from esophageal constriction or web (Fig. 2). Dermatological examination revealed koilonychia with brittle nails (Figs. 3 and 4).

The oral findings showed atrophic mucous membrane and loss of normal keratinization. The tongue appeared smooth and red. There was atrophy of the filiform papillae also (Figs. 5 and 6). Multiple root remnants were present.

A provisional diagnosis of anemic glossitis was arrived. The differential diagnosis was made with Plummer–vinson syndrome, early stage OSMF.

Blood investigation revealed hypochromic microcytic anemia, with low red blood cell count, low serum iron and ferritin level. Based upon the clinical examination and investigations the final diagnosis of Plummer–vinson syndrome was made.

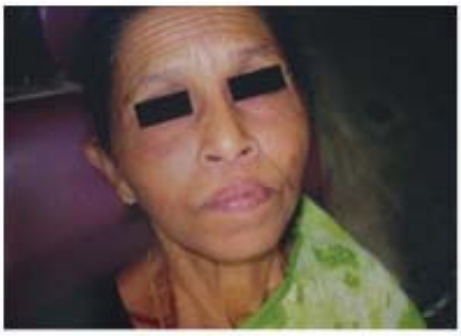

Fig. 1: Facial photograph lemon tinted pallor of skin.

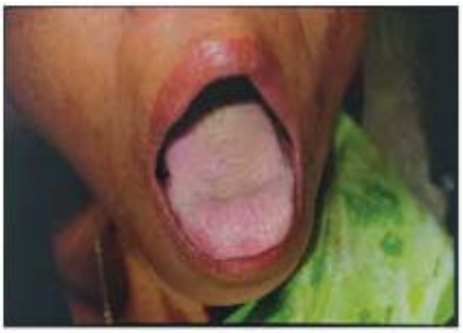

Fig. 2: Intraoral photograph showing atrophy of filiform papilla and candidiasis.

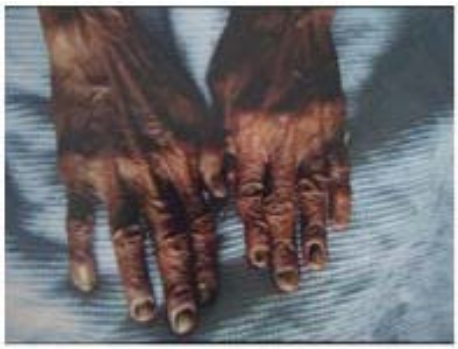

Fig. 3: Photograph of hands showing brittle nails.

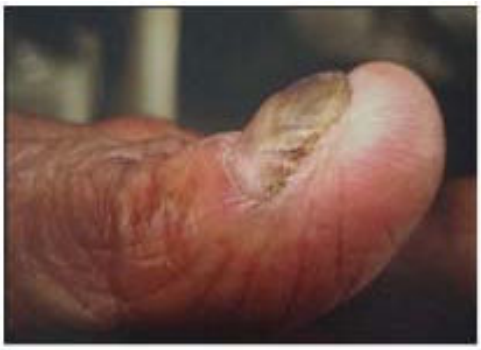

Fig. 4: Photograph of fingernail showing koilonychia.

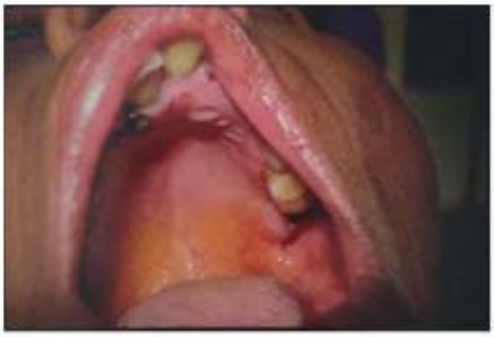

Fig. 5: Intraoral photograph showing pale areas in palate.

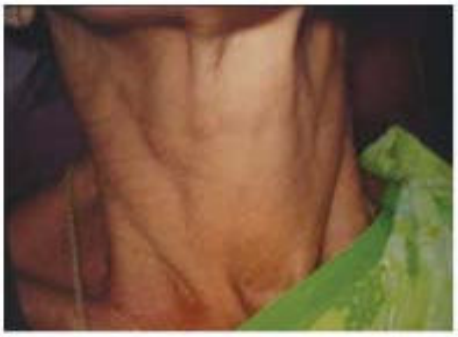

Fig. 6: Photograph of neck showing webbing.

Discussion

The exact cause of Plummer–Vinson syndrome is unknown.[2,3,5,7] The genetic factors and nutritional deficiencies may play a role in the development of this syndrome.[4,5,8–10] It is more common in women, especially in middle age and peak age over 50 years.[2–4]

Patients with Plummer–Vinson syndrome often have a burning sensation of oral cavity especially on the tongue and oral mucosa and atrophy of lingual papillae produces a smooth, shiny, red, dorsum of the tongue.[11–13] Symptoms include dysphagia (difficulty in swallowing), odynophagia (painful swallowing), atrophic glossitis and angular stomatitis.[12–14]

Serial contrast gastrointestinal radiography or upper-gastrointestinal endoscopy may reveal the web in the esophagus.[13,14] Blood tests demonstrate a hypochromic microcytic anemia that is consistent with an iron-deficiency anemia.[12–15] Biopsy of involved mucosa typically reveals epithelial atrophy. Epithelial atypia or dysplasia may be present.[16]

The dysphagia has a gradual onset, and is usually painless and intermittent. Patients usually point to the neck at or above the suprasternal notch.[9,10] Dysphagia is usually noticed first for solid foods, which is then followed after several years by difficulty in swallowing liquids. With worsening obstruction, choking and/or episodes of aspiration may occur.

Anemia can manifest with several symptoms, such as easy fatigability, exertional dyspnea, weakness and palpitations. Physical examination of patient reveals pallor.[8–11,15] It may associated with glossitis, angular cheilitis, koilonychias, seborrheic dermatitis, hyperkeratosis, conjunctivitis, keratitis, paresthesia and may be night blindness.[1–4,12–14]

The objectives of investigations in a patient with this syndrome are to diagnose anemia, ascertain the cause of anemia, assess the severity and cause of dysphagia and to localize the obstructing lesion to help plan definitive treatment. Hence, the investigations include hematologic tests, and radiographic and endoscopic examinations of the esophagus.[7,8,14–17] The further investigations may be required to identify associated conditions such as thyroid disorders, celiac disease and so on to exclude hypopharyngeal or esophageal malignancy. To identify and look for occult blood loss, endoscopic examination to look for lesions in the distal gastrointestinal tract and investigations to look for the cause of blood loss from the female genital tract is important aspect.[15–19]

Hemoglobin concentration in blood, microscopic examination of peripheral blood smear, red cell indices like mean corpuscular volume, mean corpuscular hemoglobin and mean corpuscular hemoglobin concentration and serum iron studies like serum iron, ferritin and total iron binding capacity are helpful in diagnosing the presence or absence of anemia and to establish iron deficiency as its cause[13,14,17,18] Barium swallow radiography is the investigation most commonly asked for suspected esophageal web.[15–17,19]

Treatment for dysphagia, and iron supplementation resolves disease process. This treatment alone could be considered for those with mild dysphagia or if endoscopy facilities are not available.[2,3,13,15,17–19] Advanced and long-standing dysphagia is unlikely to respond to iron replacement alone and requires dilatation of the web. Iron therapy should be considered in all patients with web, regardless of the hemoglobin status, to replenish the iron stores.[15,17,19]

It is considered to be a precancerous condition and such patients are considered to be at risk of developing squamous cell carcinoma of the hypopharynx or upper esophagus. The long-standing iron deficiency, the atrophic mucosal changes become irreversible and, in some patients, progress to malignant degeneration. [14–19]

Conclusion

Follow up of patient is necessary due to its premalignant behavior and increased incidence of malignant transformation. Future research is needed to clarify the pathogenesis, nature and treatment of Plummer–Vinson syndrome.

References

1. Plummer HS. Diffuse dilatation of the esophagus without anatomic stenosis (cardiospasm): A report of 91 cases. JAMA. 1912;58:2013–15.
2. Masri O, Sharara AI. Plummer–Vinson syndrome. Clin Gastroenterol Hepatol. 2013;11(12):85.
3. Gude D, Bansal D, Malu A. Revisiting Plummer–Vinson syndrome. Ann Med Health Sci Res. 2013;3(1):119–21.
4. Howell JT, Monto RW. Syndrome of anemia, dysphagia and glossitis (Plummer–Vinson

syndrome). N Engl J Med. 1953;249(25):1009–12.

5. Klifto EJ, Allen SK, Metzman M, et al. Plummer–Vinson syndrome: Report of a case and review of the literature. J Am Osteopath Assoc. 1983;83(1):56–59.
6. Nosher JL, Campbel WL, Seaman WB. The clinical significance of cervical esophageal and hypopharyngeal webs. Radiology. 1975;117(1):45–47.
7. Vinson P. A case of cardiospasm with dilatation and angulation of the esophagus. Med Clin North Am. 1919;3:623–27.
8. Kelly A. Spasm at the entrance of the esophagus. J Laryngol Rhinol Otol. 1919;34:285–89.
9. Paterson D. Clinical type of dysphagia. J Laryngol Rhinol Otol. 1919;34:289–91.
10. Chisholm M. The association between webs, iron and post-cricoid carcinoma. Postgrad Med J. 1974;50(582):215–19.
11. Bredenkamp JK, Castro DJ, Mickel RA. Importance of iron repletion in the management of Plummer–Vinson syndrome. Ann Otol Rhinol Laryngol. 1990;99(1):51–54.
12. Adlington P, Leedham P, Smith P. The place of iron deficiency in the Paterson–Brown–Kelly syndrome. J Laryngol Otol. 1973;87(9):845–59.
13. Jacobs A. The buccal mucosa in anaemia. J Clin Pathol. 1960;13(6):463–68.
14. Geerlings SE, Statius van Eps LW. Pathogenesis and consequences of Plummer–Vinson syndrome. Clin Investig. 1992;70(8):629–30.
15. Atmatzidis K, Papaziogas B, Pavlidis T, et al. Plummer–Vinson syndrome. Dis Esophagus. 2003;16:154–57.
16. Entwistle CC, Jacobs A. Histological findings in the Paterson–Kelly syndrome. J Clin Pathol. 1965;18(4):408–13.
17. Waldenstrom J, Kjellberg SR. The roentgenological diagnosis of sideropenic dysphagia (Plummer–Vinson syndrome). Acta Radiologica. 1939;20:618–38.
18. Lopez A, Cacoub P, Macdougall IC, Peyrin-Biroulet L. Irondeficiency anaemia. Lancet. 2016;387(10021):907–16.
19. Cook JD. Diagnosis and management of iron-deficiency anaemia. Best Pract Res Clin Haematol. 2005;18(2):319–32.