

## Plummer Vinson Syndrome- A Case Report

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### ABSTRACT:

Plummer-Vinson or Paterson-Kelly syndrome presents as a triad of, iron-deficiency anemia, dysphagia and esophageal webs. Most of the patients include white middle-aged women, in the fourth to seventh decade of life. While literature review elucidates the resolution of dysphagia in most cases with iron therapy, we discuss our case where the dysphagia was resistant to such therapy. The patient underwent endoscopic dilatation of esophageal webs which relieved the dysphagia. These patients should be followed closely as Plummer-Vinson syndrome is associated with an increased risk of squamous cell carcinoma of the pharynx and the esophagus.

**Keywords:** Anemia, Dysphagia, Esophageal and pharyngeal carcinoma

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### INTRODUCTION

The most used name is “Plummer-Vinson syndrome”, named after Henry Plummer and Porter Vinson. Another term is “Paterson-Kelly syndrome”, named after Donald Ross Paterson and Adam Brown-Kelly, who published their findings in 1919.

[1]

It is a manifestation of severe, long-term, iron deficiency anemia causing

dysphagia because of esophageal webs. The esophageal webs are thin mucosal folds protruding into the lumen of proximal esophagus.<sup>[2]</sup> <sup>1</sup> The dysphagia is usually painless and intermittent or may be progressive over the years. It may be limited to difficulty in swallowing solids initially and later liquids as well. It has been sometimes associated with weight loss.<sup>[3]</sup> Symptoms such as weakness, pallor, fatigue,

tachycardia may dominate the clinical picture. Other symptoms may include glossitis, glossopyrosis, glossodynia, angular cheilitis, koilonychia, fragility, brittle nails.<sup>[2]</sup>

Iron deficiency commonly affects females and has numerous side effects. Detailed examination of such patients is mandatory to prevent further complications. Patient counselling is also an important part as reversal of iron deficiency usually leads to regression of symptoms.

**CASE HISTORY**

A 36 year old female patient reported to the Department of Oral Medicine and Radiology, Pandit Deendayal Upadhyay Dental College, Solapur with the complaint of food lodgment in the upper right back

region since 1 month. History of endodontic treatment along with crown prosthesis was present which was done 3- 4 years back. The crown prosthesis was dislodged and misplaced by the patient 2 months back.

On further questionnaire, patient revealed a history of dysphagia 8 years back for which she had underwent endoscopy. The endoscopy showed esophageal web just below the cricopharynx for which she was prescribed hematinics. Follow up after 6 months did not regress the symptoms and therefore esophageal dilatation was done. Six years later, the patient experienced similar symptoms for which she again underwent endoscopy followed by dilatation. (Figure 1 & 2) Her social history revealed low economic status.

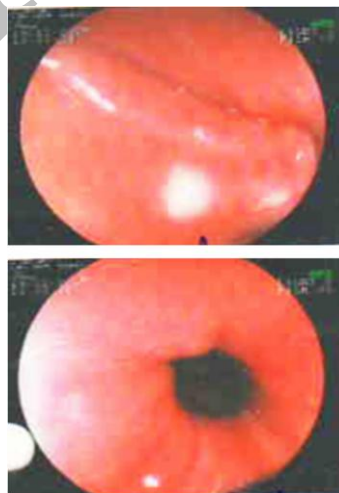


Figure 1: Esophageal webs followed by dilatation (First episode)

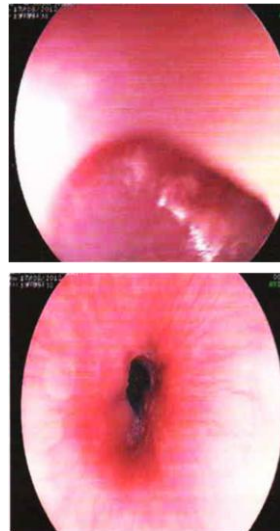


Figure 2: Second episode of esophageal dilatation.

On General physical examination, patient was poorly nourished and hyposthenic. Pallor was noted in the palpebral conjunctiva and nail beds along with flat and brittle nails. (Figure 3)



Figure 3: Brittle, flat nails and pale palpebral conjunctiva.

Intraoral examination revealed pale mucosa and bald tongue (Figure 4, 5). Angular cheilitis was also noted.



Figure 4: Pale mucosa.



Figure 5: Bald tongue.

Blood investigations revealed: Hemoglobin- 7.5g/dl, Hematocrit- 25.2%, Mean corpuscular volume- 50.70fL, Mean corpuscular hemoglobin- 15.09 pg and Mean corpuscular hemoglobin concentration- 29.76g/ dl. The RBC's were Anisocytosis +, Microcytosis ++, Hypochromia ++.

Therefore the diagnosis of "Plummer- Vinson syndrome" was established by means of medical history, clinical findings and blood investigations.

The differential diagnosis included other causes of dysphagia such as malignant tumors, benign strictures or esophageal

rings. Other reasons for dysphagia are diabetes mellitus, gastroesophageal reflux disease scleroderma, diverticula, achalasia and neuromuscular disorders. The patient was asymptomatic at the time of presentation. The patient is under follow- up and no recurrence of dysphagia has been noted until now.

### **DISCUSSION**

Etiopathogenesis of Plummer- Vinson syndrome is unknown. The most important possible etiological factor is iron deficiency. Other possible factors include malnutrition, genetic predisposition or

autoimmune processes. The deficiency of the iron dependent enzymes and high cell turnover rate in the epithelium of the upper digestive tract makes it vulnerable to iron deficiency. These of the oxidative enzymes, stress induced by free radicals and DNA damage may be responsible for the epithelial changes.

This is also predisposes to malignant transformation chiefly squamous cell carcinomas. Iron deficiency is believed to decrease the contraction amplitude of the esophageal muscle which results in impaired function and motility.<sup>[1]</sup>

The hypothesis that iron deficiency anemia may contribute to esophageal webs may also apply in our case as an etiologic factor initially. But our case being chronic long standing anemia, was refractory to iron therapy.

Very few cases have been reported that have been recalcitrant to iron therapy. In such circumstances, dysphagia should be treated by mechanical dilatation in adjunction with iron therapy. In most cases, one session is usually enough for long term relief. Multiple sessions may also be

required, although rarely as reported in our case.

Logan reported that patients often tolerate progressive dysphagia for a considerable period without seeking medical attention, thus leading to late presentation.<sup>[4]</sup> This may explain why our patient did not seek medical attention for a long time.

Uygur- Bayramicli in 1999 reported esophageal strictures instead of webs, in association with iron- deficiency anemia and dysphagia.<sup>[5]</sup>

Lawoyin et al in 2008, reported a case of a mentally retarded female from Saudi Arabia with dysphagia and poorly differentiated squamous cell carcinoma of the tongue. Her blood investigations suggested anemia for which she was hospitalized and treated. The patient eventually expired due to worsening pneumonia and cardiopulmonary arrest.<sup>[8]</sup>

Sudhir Naik et al in 2011 reported a case of a female with easy fatigue, dehydration and dysphagia. Dilatation of esophageal webs was done by cuffed endotracheal tube after which the dysphagia improved.<sup>[9]</sup>

A upper gastrointestinal endoscopy, barium swallow study are usually advised for the detection of a web, although the best method is the videofluoroscopy. [1,6] Mercury/tungsten filled bougies, bougienage dilators and balloon dilators are the esophageal dilators commonly available.[7] The prognosis is good, although patients should be followed up regularly for the possibility of malignant transformation.

**CONCLUSION**

This patient was asymptomatic during presentation and did not reveal her past medical history during initial questionnaire. Therefore, detailed case history with main focus on the general clinical examination of the patient is mandatory as it can lead to important clues to the diagnosis. Since it is known that patients with Plummer- Vinson syndrome are more prone to develop esophageal and oral cancer, it is important to investigate in depth and follow up such patients.

**REFERENCES**

1. Gottfried Novacek. Plummer-Vinson syndrome. *Orphanet J Rare Dis.* 2006; 1: 36.

2. Gude, et al.: Understanding plummer vinson syndrome. *Annals of Medical and Health Sciences Research*, 2013 .Vol 3.

3. Godina J, Wong PW, A triad of trouble finding: Plummer Vinson Syndrome. *Postgrad Med.*2000. 108; 109-10.

4. Logan JS, A review of the Plummer-Vinson Stricture of the cervical esophagus. *Ulster Med J.* 47, Suppl 2; 1-14.

5. Uygur- Bayramicli O, Tuncer K, Dolapcioglu C. Plummer Vinson syndrome presenting with an esophageal stricture. *J Clin Gastroenterol* 1999; 29(3); 291-92.

6. Layowin Davidson O. *Internet Journal of Dental Science.* 2008, Vol. 5, p6-6.

7. Sudhir M Naik, Shivakumar MC, Mohan Appaji. *International Journal of Head and Neck Surgery.* 2011; 2(3): 161-165.

8. Dysphagia in a Patient with RA and iron deficiency anemia. *Medscape*

*General Medicine* 4 (3), 2002. @  
2002 Medscape.

9. Egan JV, Baron TH, Adler DG,  
Davila R, Faigel DO, Gan SL, *et al.*;

Standards of practice committee.  
Esophageal dilation. *Gastrointest*  
*Endosc* 2006;63: 755-60.

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