Case Report

**Plummer-Vinson Syndrome**
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**Abstract**
Plummer-Vinson or Paterson-Kelly syndrome presents as a classical triad of dysphagia, iron deficiency anemia and esophageal webs. It typically affects middle aged females who may have other features of chronic iron deficiency, namely papillary atrophy of the tongue, spoon shaped brittle nails, angular stomatitis and pica. Since the dysphagia is associated with iron deficiency anaemia, the term sideropenic dysphagia has often been used to describe this condition. The oesophageal web is best diagnosed on barium swallow. Etio-pathogenesis of Plummer-Vinson syndrome is unknown but the most important possible etiological factor is iron deficiency. Other possible factors include malnutrition, genetic predisposition or autoimmune processes. Plummer-Vinson syndrome can be treated effectively with iron supplementation. Since Plummer-Vinson syndrome is associated with an increased risk of squamous cell carcinoma of the pharynx and the esophagus, the patients should be followed closely. The incidence of this condition is decreasing; however, because of better nutrition and improved health care. Since PVS is associated with an increased risk of squamous cell carcinoma of the pharynx and the esophagus, the patients should be followed closely with GI endoscopic examination at regular interval. This paper presents a case of a 69-year-old woman with Plummer–Vinson syndrome who was successfully treated.

**Keywords**

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Introduction

Plummer Vinson syndrome (also known as Paterson-Kelly syndrome, Paterson-Brown-Kelly Syndrome, Sideropenic Dysphagia) is defined as the classical triad of dysphagia, iron deficiency anemia and oesophageal webs. It was first described in 1912, common in middle aged females, in fourth to seventh decade of life [1, 2]. It is characterized by dysphagia, usually due to upper oesophageal webs, anemia and other features like angular chelitis and glossitis and koilonychia and also other anemia related features like weakness, pallor, fatigue and tachycardia. Enlargement of spleen and thyroid may also be seen in some cases. Dysphagia is usually painless and intermittent or progressive over the years, limited to solids, sometimes associated with weight loss [2].

The most accepted etiological factor behind Plummer Vinson Syndrome is iron deficiency and other possible factors include malnutrition, genetic predisposition or autoimmune process [3]. The diagnosis is based on the evidence of iron deficiency anemia and one or more oesophageal webs which can be detected by barium swallow X-ray, but the best way for demonstration of webs is videofluoroscopy [2, 4]. Webs can also be demonstrated by gastrointestinal endoscopy. Iron supplementation alone in these types of cases can resolve dysphagia in many patients [2].

Plummer Vinson syndrome can be treated effectively with iron supplementation and mechanical dilation of webs. Plummer Vinson syndrome has been identified as a risk factor for the development of squamous cell carcinoma of upper gastrointestinal tract, which accounts 3 to 15 % of the cases [5, 6]. Here we are reporting a case of a 69 year old woman with features of Plummer Vinson syndrome.

Case Report

Case presentation

A 69 years old female patient reported to the department Oral Medicine, Diagnosis, and Radiology, Government Dental College and Hospital, Mumbai with the chief complaint of difficulty in swallowing and burning sensation in the mouth since last 6 months. The patient felt discomfort in her lower part of the neck when swallowing solid food but no pain was reported as such. Difficulty in swallowing was not associated with any change in posture or other symptoms and continued to be the same till date without much aggravation or relief. The patient also started having a burning sensation in the mouth since last 2 months before reporting to our department. On further questioning it was revealed that the burning was present throughout the mouth and aggravated by spicy foods. The patient reported no change in her salivation. There was no history of malena.

On her past medical history the patient gave history of breathlessness since 3-4 months and there was no evidence of any other serious illness before this problem. Patient was not taking any kind of medication and had no any abusive habits of paan masala, supari or gutkha chewing.

During examination the patient was thin and had low body weight. The skin of the patient was rough and dry and her hairs were also rough, untidy and kinky. The nails of hands and legs were brittle and abnormal in appearance (Figure 1).

Intraoral findings suggested bilateral atrophic, blanched, pale oral mucosa, depapilated surface of tongue (Figure 2A). Mouth opening of the patient was limited and angular cheilitis was present. Pallor or paleness was present on palpebral conjunctiva (Figure 2B).
Investigations

Haematological investigations performed as an outpatient, showed reticulocyte count: 4.8% (normal: up to 1%), haemoglobin level: 6 gm% (normal: 12-16 gm%), iron level: 21μg/dl (normal: 37-145μg/dl), ferritin level: 2.86 ng/ml (13-150ng/ml), transferrin saturation: 6.55% (13-45%), TIBC: 320.40μg/dl (228-42840μg/dl). Peripheral smears showed anisocytosis, poikilocytosis, microcytosis, and hypochromasia.

There was no evidence of other cause of iron deficiency anemia such as malabsorption, malnutrition, use of NSAIDs and abnormal vaginal bleeding.

Esophagography disclosed an oesophageal web on left side opposite to C6 vertebra and smooth short segment narrowing of upper part of the oesophagus opposite to C6 and C7 vertebra level (Figure 3).

Differential diagnosis

The above findings, along with upper esophageal webs, iron deficiency anemia and glossitis, was suggestive of the diagnosis of Plummer–Vinson syndrome.

Treatment
The patient received 40 mg of intravenous iron therapy daily for 30 days.

**Outcome and follow-up**

The anemia and dysphagia thereby improved and the patient was kept under follow up.

**Discussion**

High incidence of Plummer Vinson syndrome in females can be attributed to inadequate dietary intake, chronic blood loss in form of menstruation [7]. The exact data about the incidence and prevalence of Plummer Vinson syndrome is not known, however the incidence has reduced recently because of the nutritional improvement, advanced health care, and better treatment of iron deficiency [8, 3]. The proposed pathogenesis behind the various clinical manifestations is the reduction of iron dependent enzymes which may cause mucosal degeneration, atrophic changes and web formation, further leading to cancer development [9, 10]. The most possible etiology is iron deficiency, however other factors also, such as genetic predisposition, malnutrition and autoimmune conditions have been found to play a role in the pathogenesis of this syndrome [11].

In our case malnutrition leading to iron deficiency can be the main etiology of the disease. The characteristic clinical features may include glossitis, glossopyrosis, glossodynia, angular cheilitis, koilonychia, fragility, thinning of nails, and brittle hair. Rare manifestations such as clubbing instead of koilonychia and tortuous esophagus in addition esophageal webs have also been described. Symptoms secondary to anemia such as pallor, fatigue, and weakness may also dominate the clinical picture [13].

The dysphagia associated with Plummer Vinson syndrome is usually painless and intermittent or progressive over the years, limited to solids and sometimes associated with weight loss [12]. Grade I dysphagia is where in patient has only difficulty in swallowing solids, Grade II dysphagia is where in patient can take only semisolids, Grade III dysphagia is where in patient can take only liquid and difficulty to swallow even saliva is Grade IV [7]. Our patient fell into the category of Grade I dysphagia.

Differential diagnosis includes all other conditions which can be associated with dysphagia such as malignant tumors, benign strictures or esophageal rings, various other reasons for dysphagia can be diverticula, motility disorders such as achalasia, spastic motility disorders, scleroderma, diabetes mellitus, gastroesophageal reflux disease and neuromuscular skeletal muscle disorders [3].

In our case laboratory investigations revealed increased reticulocyte count, decreased haemoglobin level, iron level, ferritin level, transferrin saturation and peripheral smears showed anisocytosis, poikilocytosis, microcytosis, and hypochromasia.

All the above lab findings were characteristic of iron deficiency anemia and we also ruled out other causes of dysphagia. In this case barium swallow also revealed stricture formation in proximal part of oesophagus, hence we arrived at the diagnosis of Plummer Vinson syndrome secondary to iron deficiency anemia. This syndrome is associated with an increase incidence of postcricoid carcinoma (3-15%), so it is considered as a precancerous lesion [5] and in rare cases of the stomach also [14, 15].

This disease can be treated effectively with iron supplementation, which can resolve dysphagia in most of the cases, but before starting the treatment the other causes of iron deficiency such as active hemorrhage, malignancy or celiac disease should be taken into consideration and ruled out. However, in cases of significant obstruction of the esophageal lumen by the webs and persistent dysphagia despite iron
supplementation, rupture and dilation of the web should be performed [3]. Apart from endoscopic dilatation, argon plasma coagulation therapy of esophageal webs has been tried with reasonable success [16]. Prognosis of Plummer Vinson syndrome is excellent as dysphagia and anemia can be treated effectively, but in cases of associated carcinoma of hypopharynx or upper oesophagus, the prognosis worsens.

It is important that this syndrome be differentiated from other causes of dysphagia, e.g. malignant tumors, strictures, esophageal burns, heterotopic gastric mucosa or blistering skin disease. Even though a barium swallow is the method of choice for diagnosis of Plummer Vinson syndrome, a careful upper GI endoscopy with introduction of the endoscope under vision past the cricopharyngeus, is better as it allows for therapy via endoscopic dilatation which is successful in most cases. Since PVS is associated with an increased risk of squamous cell carcinoma of the pharynx and the oesophagus, the patients should be followed closely with GI endoscopic examination at regular interval. Biopsy of any suspicious lesion on endoscopy and regular follow up is essential.

**Conclusion**

Future research is needed to clarify the pathogenesis, nature and treatment of Plummer Vinson syndrome.

**References**

12. Rania Hefaiadh et al, Plummer-Vinson syndrome, La Tunisie Medicale 2010; Vol 88 (n°10) : 721 – 724