

# A Case of Plummer-Vinson Syndrome Esophageal Web Dysphagia treated by Dilatation with Cuffed Endotracheal Tube

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## ABSTRACT

**Background/objectives:** Plummer-Vinson syndrome also known as sideropenic dysphagia is a disease characterized by chronic iron-deficiency anemia, dysphagia and esophageal web. It commonly affects white female in the 4th to 7th decade. Most of the dysphagia and iron deficiency can be treated by iron supplementation and rarely web dilatation is needed.

**Setting:** Department of ENT, Head and Neck Surgery and Anesthesia, KVG Medical College, Sullia, Karnataka, India.

**Case report:** A 36-year-old female with dysphagia of 10 months and iron-deficiency anemia with a small upper esophageal web seen on upper GI endoscopy and barium swallow.

**Intervention:** Conservative line of management with blood transfusion and dilatation of the web with cuffed endotracheal tube.

**Results:** A good symptomatic and radiological improvement was seen after blood transfusion and web dilatation with cuffed endotracheal tube.

**Conclusion:** Cuffed endotracheal tube dilatation is a better way of managing upper esophageal webs with minimal complications under general anesthesia.

**Keywords:** Plummer-Vinson syndrome, Dysphagia, Iron-deficiency anemia, Koilonychia, Carcinoma.

## INTRODUCTION

Plummer-Vinson syndrome (PVS) was first described by Patterson and Kelly in 1919.<sup>1</sup> The syndrome consist of dysphagia, atrophic oral mucosa, glossitis and anemia and most of the patients affected are postmenopausal women.<sup>2</sup> The triad consists of dysphagia, iron-deficiency anemia and esophageal webs.<sup>3</sup>

This can be treated effectively with iron supplementation and if necessary by dilatation by webs.<sup>3</sup> The disease is rare nowadays, but its important because it identifies the risk of squamous cell carcinoma of the postericoid pharynx and the upper esophageal region.<sup>3</sup> Other presenting symptoms may include cracks or fissure at the corners of the mouth along with painful tongue.<sup>4,5</sup> Koilonychia (spoon-shaped finger nails) or nails that are brittle which break easily and other classical features of iron deficiency are evidently seen.<sup>4,5</sup>

In approximately 10% of cases, this uncommon syndrome is associated with hypopharyngeal or esophageal cancers and rarely oral cavity carcinoma which arises from the degenerative changes in the mucosae of the pharynx, esophagus and oral cavity rarely.<sup>6-8</sup> Here, we report a case of dilatation of the esophageal web done using the cuffed

endotracheal tube used for microlaryngeal surgery under general anesthesia along with the clinical features of this rare syndrome.

## CASE REPORT

A 36-year-old female presented with difficulty in swallowing since 10 months duration. The patient was a short stratured female of 135 cm. The patient gave history of easy fatiguability and physically looked emaciated, severely dehydrated, pale and lethargic.

She weighed 35 kg and her temperature (oral) was 39.8°C. Blood pressure was 100/60 mm Hg, pulse 98/minute and respiration was 22/minute. There was bilateral angular stomatitis with epithelial crust on the lips and bald tongue. The skin appeared generally dry and the finger nails of hands and feet were spoon shaped. Chest auscultation revealed a normal heart sounds and minimal bilateral basal crepitation in the lungs.

The abdomen was soft, nontender and liver, spleen and kidneys were not palpably enlarged. Intraorally, the oral mucosa was dry with blanched mucosa all round and bald tongue with absent papillae. Hematologic parameters



Fig. 1: Glossitis, bald tongue and cheilitis seen



Fig. 4: Koilonychia seen in fingers



Fig. 2: Koilonychia of foot



Fig. 5: Barium swallow oblique showing narrowing



Fig. 3: Koilonychia of fingers



Fig. 6: Barium swallow AP view showing narrowing

revealed a WBC of 4800 cells/mm<sup>3</sup>, hemoglobin 7.4%, neutrophils 57%, lymphocytes 36%, eosinophils 2%, monocytes 5%, ESR 11 mm/hr, AEC 100 cells/mm<sup>3</sup> and platelet count 5.6 lakhs.

The peripheral blood smear showed RBCs to vary from dimorphic to normocytic normochromic cells. Most of them were microcytic hypochromic cells with few macrocytes and polychromatophilic cells. WBCs were normal in number,



Fig. 7: Endoscopic view of the partial web



Fig. 10: AP view barium swallow after dilatation

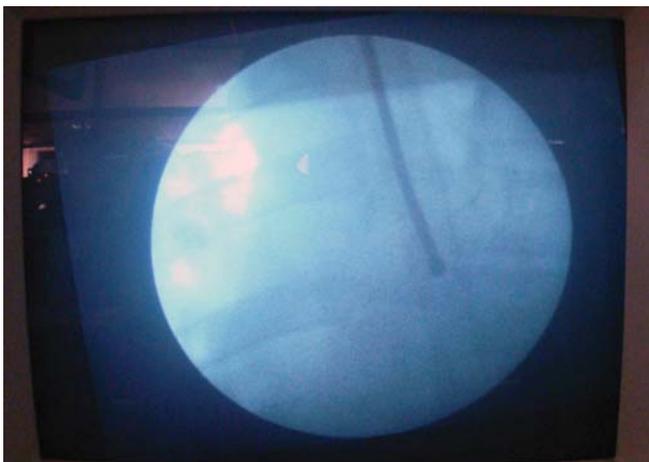


Fig. 8: Cuffed ET tube dilatation of web being done

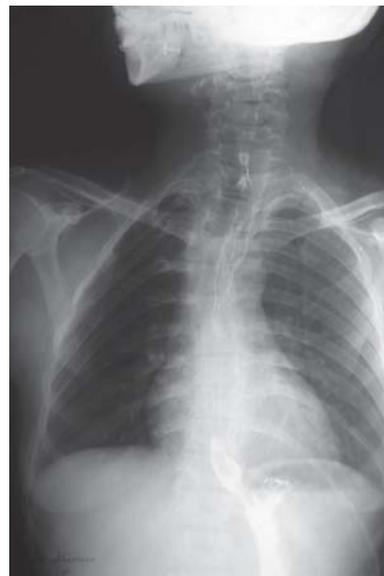


Fig. 11: Oblique view barium swallow after dilatation



Fig. 9: Web dilatation done with C-arm under general anesthesia

morphology and distribution but a mild increase in platelets was seen. Overall a dimorphic anemia with mild thrombocytosis was seen. The blood group was B +ve with normal bleeding and clotting time. T3 was 1.23 ng/ml, T4 was

8.4 micro gm/dl and TSH was 2.63 micro IU/ml. HIV 1, 2 and HBsAg ELISA were negative on examination. Chest X-ray showed scattered bronchopneumonic changes in the mid- and lower zones.

After admitting the patient blood transfusion was planned and 3 pints of blood were transfused at 2 days interval. The Hb% was monitored along with the blood transfusion and the progressive improvement in dysphagia assessed. Upper GI endoscopy was done and mild web-like condition was seen in the upper esophagus and rest of the esophageal and gastric mucosa was normal. Barium swallow was done and the narrowing confirmed at the upper esophagus.

As dysphagia did not improve, dilatation of the web under general anesthesia was planned using dilatation by cuffed endotracheal tube. Dilatation was done by cuffed

long endotracheal tube used for microlaryngeal surgery. The dilatation was monitored radiologically using C-arm. The patient had an uneventful recovery after the procedure and dysphagia improved.

She complained of some pain while swallowing on the operative day and it subsided later on. The dysphagia subsided and was better than that it was preoperatively. The patient was discharged on the 4th postoperative day and regular follow-up with endoscopy at 2 months interval done for one year.

## DISCUSSION

PVS was described as a symptom complex in 1919 which included dysphagia, iron deficiency and presence of superior esophageal web.<sup>9,10</sup> The syndrome mainly affects white women, in the 4th to 7th decade of life, but some pediatric and adolescent cases are also reported.<sup>9-12</sup>

The disease is rarer nowadays and the incidence is decreasing even in African nations where iron deficiency and malnutrition are common.<sup>13</sup> The incidence and prevalence are not reliable as hematological parameters are not included.<sup>14</sup>

The pathogenesis of the syndrome is unclear but iron-deficiency anemia, malnutrition, genetic predisposition and autoimmune etiologies are postulated.<sup>10,15</sup> Iron-deficiency anemia is the widely accepted etiology as dysphagia and esophageal webs improve with iron supplementation.<sup>10</sup> Upper esophageal webs are seen in only 10% of patients with iron deficiency which denotes a multiple etiologies in the syndrome.<sup>16</sup> Iron deficiency causes reduction of iron-dependent oxidative enzymes which results in gradual degradation of the muscles of the pharynx leading onto mucosal atrophy and development of webs.<sup>15</sup>

Whatever be the cause of the iron deficiency, the theory of web formation is based on the rapid losses of iron-dependent enzymes due to its high cell turnover.<sup>12,17</sup> Reduction of these enzymes may cause mucosal degenerations, atrophic changes and web formation which causes neoplastic changes in the lower pharynx and upper esophagus.<sup>12,17</sup>

Also iron deficiency decreases the rate esophageal muscle contraction amplitude leading to motility impairment.<sup>18,19</sup> The webs limited to the proximal and middle part of the esophagus can be explained on the finding that the transit time is slower in the upper esophagus in iron deficient individuals compared with normal volunteers.<sup>20</sup>

The patients usually complain of dysphagia to solids, but the symptoms can progress to dysphagia to liquids as well.<sup>11</sup> The dysphagia is usually nonpainful, leading onto progressive weight loss.<sup>10,21,22</sup> As the progressive dysphagia is painless, patients tolerate it for long before presenting to the clinician.<sup>23</sup> Symptoms of iron-deficiency anemia like pallor, fatigue and weakness dominate the clinical picture along with dysphagia.<sup>24</sup>

The syndrome has a risk of squamous cell carcinoma of the postcricoid pharynx and upper esophagus.<sup>2</sup> Around 3 to 15% of the patients mostly women between 15 and 50 years of age, have been reported to develop esophageal or pharyngeal cancer.<sup>25,26</sup> The syndrome is proven to be precancerous as 10% of the neoplastic changes in the hypopharynx, oral cavity and upper esophagus are malignant squamous cell carcinomas.<sup>27</sup> A rare association of base of the tongue cancers have been associated with this syndrome in Scandinavian countries.<sup>27</sup>

Uygun-Bayramicli et al reported cases presenting with upper esophageal stricture instead of web in some of the cases with dysphagia and iron-deficiency anemia.<sup>28</sup> Other causes of dysphagia like malignant tumors, strictures, esophageal burns and heterotopic gastric mucosae have to be ruled out.<sup>29,30</sup> The patients should be followed up by upper GI endoscopies as neoplastic changes with the webs are seen.<sup>29,30</sup> Barium swallow is investigation of choice to detect these esophageal webs and upper GI endoscopies are confirmatory in diagnosis of this condition.<sup>10,12</sup>

Management includes diagnosing the cause of anemia like active hemorrhage, malignancy or celiac disease.<sup>10</sup> The syndrome can easily be treated by iron therapy and the webs by mechanical dilatation.<sup>10</sup> Iron supplementation can resolve dysphagia in many patients without mechanical dilatation.<sup>10</sup> Iron therapy is advised even if the hemoglobin percentage is normal in the presence of web formation.<sup>18</sup> Patients with choking and aspirations need dilatation therapy along with iron supplementation.<sup>31,32</sup>

The prognosis of this condition is good as anemia and dysphagia can be effectively treated by iron therapy and the webs by dilatation.<sup>2</sup> The prognosis worsens dramatically if the syndrome is associated with complications like squamous cell carcinomas of hypopharynx and upper esophagus.<sup>10</sup> Endoscopic dilatation is the procedure of choice for treating esophageal webs.<sup>31,32</sup>

In our case, dilatation of the esophageal web was done using cuffed endotracheal tube under general anesthesia.<sup>33</sup> This is the first time reported in literature where esophageal web dilatation using cuffed endotracheal tube has been done although balloon dilatation has already been reported.<sup>33</sup> The patient had good symptomatic and radiological improvement after therapy.

## CONCLUSION

Dysphagia and anemia of the syndrome can be treated effectively by iron supplementation under medical line of treatment giving good prognosis. But an associated squamous cell carcinoma of the hypopharynx or upper esophagus worsens the prognosis dramatically.

Here, we have described a unique way of dilatation of the upper esophageal web using the cuffed endotracheal tube after iron supplementation. The prognosis is good but

due to the possibility of malignant transformation, regular follow-up is necessary.

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