A CASE REPORT OF PLUMMER-VINSON SYNDROME

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ABSTRACT
Plummer Vinson syndrome (PVS) is a rare syndrome (also known as Paterson-Kelly syndrome, Paterson-Brown Kelly Syndrome, Sideropenic Dysphagia) is characterized by classical triad of dysphagia, iron deficiency anemia and Oesophageal webs. A 18 years female patient was admitted in General medicine Department, with chief complaints of dysphagia (difficulty in swallowing) of both liquids and solids since 1month. On enquiry she told that, when she eats solids she feels the food is stuck in the throat. Upper Gastro intestine endoscopy shows post cricoid web and Ultra sound scan (USG) of abdomen revealed Coarsened hepatic echo texture and mild splenomegaly. This is known to be associated with an increased risk of upper GI tract cancers. So they will need regular follow up and surveillance upper gastro intestinal endoscopy every year. Future research is needed to clarify the etiology & Pathogenesis.

Key words: Plummer Vinson Syndrome, Dysphagia, Iron Deficiency Anemia, Oesophageal cricoids

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INTRODUCTION:

Plummer Vinson syndrome (PVS) is a rare syndrome (also known as Paterson-Kelly syndrome, Paterson-Brown Kelly Syndrome, Sideropenic Dysphagia) is characterized by classical triad of dysphagia, iron deficiency anemia and oesophageal webs [1]. Oral characteristics include glossitis, glossopyrosis, and angular cheilitis [2]. Its etiology is unknown although autoimmune, genetic, infectious and nutritional factors have been proposed as a cause [3]. Dysphagia is usually painless and intermittent or progressive over the years, limited to solids, sometimes associated with weight loss [4]. In addition to the main symptoms, viz., hypochromic anemia, angular stomatitis, early loss of teeth, keratitis, koilonychias and visual disturbances will take place in severe condition of Plummer Vinson syndrome [5].

The pathogenesis of dysphagia is not clearly understood, both mechanical obstruction and oesophageal dysmotility have been implicated. There is evidence that iron deficiency leads to decrease in the concentration of skeletal muscle myoglobin. Manometry has demonstrated a low swallowing pressure at the pharyngeal level and low-amplitude contractions in the oesophageal region. Iron replacement has been shown to increase the amplitude of contraction and also improve dysphagia [6]. When a patient with Iron deficiency anaemia complains of difficulty in swallowing, then one should suspect Plummer Vinson Syndrome and should be investigated for the presence of upper oesophageal web [3]. Upper GI endoscopy is the safe and economical way of finding the cause of dysphagia. If there is any oesophageal web the therapeutic intervention of mechanically dilating the oesophageal web can be carried out in the same setting [4]. Usually dysphagia will be relieved after single dilatation, rarely multiple dilation is required [6]. Identification of oesophageal web in these patients should prompt us to keep these patients under surveillance for squamous cell Carcinoma of the upper alimentary tract [7, 8].

CASE PRESENTATION

A 18 years female patient was admitted in General medicine Department, with chief complaints of dysphagia (difficulty in swallowing) of both liquids and solids since 1 month. On enquiry she told that, when she eats solids she feels the food is stuck in the throat. On further questioning by the physician, it was revealed that the burning was present throughout the mouth and aggravated by spicy foods. The patient reported no change in her salivation.
On her past medical history, 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 and there was no History of malena and weight loss.

On General Physical Examination, she was Pallor (++), facial puffiness, signs of hyperdynamic circulation, Blood pressure (BP) was low-100/70mmHg, Cardiovascular sounds and per-abdomen were found to be normal at diagnosis (NAD).

**Investigations:** Hematological investigations performed as an outpatient, showed reticulocyte count: 4.8% (normal: up to 1%), hemoglobin level: 6 gm% (normal: 12-16 gm%), iron level: 21μg/dl (normal: 37- 145μg/dl), Peripheral smears showed microcytosis, and hypochromasia. White blood cells (WBC): 5000 cells/cumm (4500-11000), Polymorphs:50% (60-75), Lymphocytes:45% (20-25), Eosinophils:2% (1-5), Monocytes: 3% (1-2), Random blood sugar, Blood urea nitrogen, Serum creatinine, Serum proteins and Liver function tests were found to be normal.

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

**Differential diagnosis:** Upper Gastro intestine endoscopy shows post cricoid web and Ultra sound scan (USG) of abdomen revealed Coarsened hepatic echo texture and mild splenomegaly. The above findings, along with upper oesophageal webs, iron deficiency anemia was suggestive of the diagnosis of SEVERE ANEMIA with POST CRICOID WEB (PLUMMER VINSON SYNDROME), shown in picture no 1.

**Treatment:** The patient was received Tab.IFA (Iron + folic acid)/OD, Tab.B Complex /OD, Tab.Cefixime 200mg/BD, Inj.Optineuron (Multivitamin)lamp/IM/OD, 2 units of packed cell transfusion of same therapy for 6 days. Then she was discharged with the following medications, Tab.Folvit/OD, Tab.Multivitamin/OD, Tab.Pantoprazole 40mg /OD, Tab.Livogen (Iron + folic acid)/OD for 15 days.

**Outcome and follow-up:** The anemia and dysphagia thereby improved and the patient was kept under follow up.
DISCUSSION

Plummer-Vinson syndrome eponym has been frequently discussed. The most used name is Plummer-Vinson syndrome, named after Henry Stanley Plummer (1874–1936) and Porter Paisley Vinson (1890–1959) who were physicians on the staff of the Mayo Clinic. Another term is Paterson-Kelly syndrome, named after, Donald Ross Paterson (1863–1939) and Adam Brown-Kelly (1865–1941), both British laryngologists, who published their findings independently in 1919. They were the first to describe the characteristic clinical features of the syndrome \[^9\]. PVS has been described to have a prevalence of <1/1,000,000 [9]. Middle aged females form the majority of the cases with the ratio of male to female being reported 4:1 or higher. This disease is also repeated to have a higher incidence amongst Europeans. A number of studies also describe the incidence of PVS in children and adolescents but it is a rare presentation [10, 11].

Diagnosis is established by confirmation of iron deficiency anemia with demonstration of web in barium studies, video fluoroscopy or upper GI endoscopy \[^9\].
Table 1: Investigations to confirm Plummer Vinson syndrome

<table>
<thead>
<tr>
<th>S. no</th>
<th>Investigations</th>
<th>In this case</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Hemoglobin (g/dL)</td>
<td>6 gm% (normal: 12-16)</td>
</tr>
<tr>
<td>2</td>
<td>Peripheral blood film</td>
<td>Hypochromic &amp; Microcytic</td>
</tr>
<tr>
<td>3</td>
<td>Serum iron (mg/dL)</td>
<td>21μg/dl (normal: 37-145)</td>
</tr>
<tr>
<td>4</td>
<td>Transferrin saturation (%)</td>
<td>Not advised</td>
</tr>
<tr>
<td>5</td>
<td>Upper GI endoscopy</td>
<td>Post cricoid web</td>
</tr>
<tr>
<td>6</td>
<td>Barium study</td>
<td>Advised</td>
</tr>
<tr>
<td>7</td>
<td>Video fluoroscopy</td>
<td>Not advised</td>
</tr>
</tbody>
</table>

Above mentioned are following investigations to diagnose and confirm the Plummer Vinson syndrome. In our case based on severity of patient condition maximum investigations, done based on patient’s economic status.

CONCLUSION

Dilatation of the esophageal cricoids web and Iron supplementation had, relieved dysphagia in this patient with Plummer Vinson syndrome and asked to review regularly. This is known to be associated with an increased risk of upper GI tract cancers. So they will need regular follow up and surveillance upper gastro intestinal endoscopy every year. Future research is needed to clarify the etiology & Pathogenesis.

REFERENCES


