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Case Report

Plummer-Vinson syndrome: a case report

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ABSTRACT

Plummer-Vinson syndrome (PVS) also named as Patterson-Brown-Kelly syndrome is a combined presentation of three things- dysphagia, iron deficiency anemia, and esophageal webs, seen more often in middle aged females. A 30-year-old female presented to us with shortness of breath on exertion and long standing dysphagia and weight loss. After investigations she was found to be severely anemic. Upper GI endoscopy revealed esophageal web. Dilatation of esophageal web was done, anemia was corrected. Patient is on regular follow-up with marked improvement in terms of weight gain and increased functional capacity.

Keywords: Anaemia, Dysphagia, Oesophageal webs

INTRODUCTION

Plummer–Vinson syndrome (PVS) also known as Paterson-Brown Kelly syndrome is an the association of iron deficiency anaemia with koilonychia, angular stomatitis, glossitis and atrophy of oesophageal mucosa in the post cricoid region,which forms and obstructing web. First described in 1912 by Henry Stanley Plummer, this disease is more prevalent among middle aged females.[1] The disease usually presents either with symptoms of anemia i.e. fatigue, shortness of breath, palpitations and progressive dysphagia. Dysphagia is usually due to upper esophageal webs and mostly for solid foods. Iron therapy may reverse the process, but there is a risk of malignant change in about 10% of the affected individuals, so they need follow up.

CASE REPORT

A 30-year-old female presented to our hospital with complaints of difficulty to swallow solid food since the past two years, shortness of breath on moderate exertion and off and on episodes of palpitations. As per patient, symptoms progressed gradually to present level when she was able to take only soft diet and liquids and it takes her more time to finish the meal. Dysphagia was with solid foods only. The patient had also noted a gradual decline in her health over the past two years. Examination revealed a rundown, middle-aged female of height 155cm, weight 43kg and body mass index of 17.9kg/m^2. Patient had pallor with ridging of nails (Figure 1) and a bald pigmented tongue (Figure 2).

The evidence of malnutrition (Figure 3) like muscle wasting was noted. Investigations revealed hemoglobin of 7.8g/dl, MCV 61.3, MCH 16.7. Blood smear showed a hypochromic microcytic blood picture. Serum iron was low at 10µg/dl, serum ferritin 3.8, iron binding capacity of 332µg/dl and transferrin saturation of 8.6%. Vitamin Band folate levels were normal. None of the other lab tests showed any abnormalities. Upper gastrointestinal (GI) endoscopy showed a narrowing in the upper esophagus confirmed to be an esophageal web (Figure 4) about 14cm from the incisor. No other abnormality was seen. Webotomy was done during the procedure. Based on the above findings, a diagnosis of PVS was made.

Following 6 hours of procedure, patient was able to take diet without much dysphagia. Patient is on regular follow up and has marked improvement in signs, symptoms and overall wellbeing.

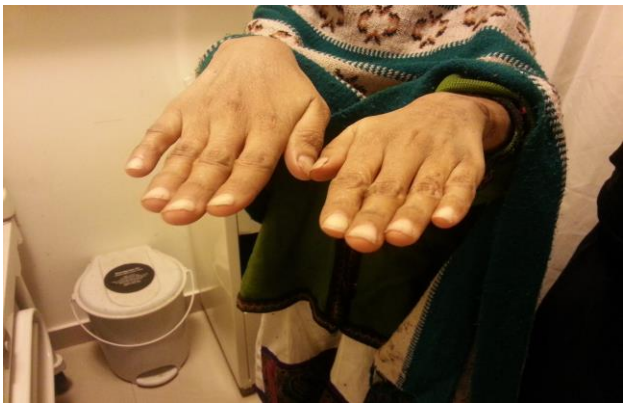

Figure 1: Patient nails depicting ridging (koilonychias).

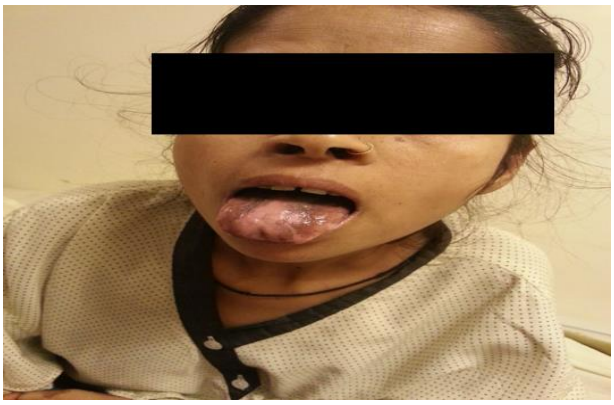

Figure 2: Patient having bald and pigmented tongue.

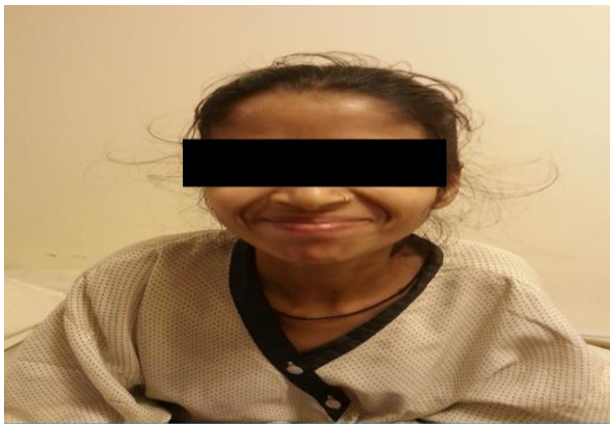

Figure 3: Patient having malnutrition.

DISCUSSION

PVS is a very rear disease. Prevalence has been around <1/1,000,000. Mostly, middle aged females[1]are affected. The patients usually present with features of anemia like fatigue, loss of appetite, weakness, palpitation, dyspnea on exertion, dizziness and dysphagia also develops. The dysphagia is progressive over years, usually to solids initially which may be associated with weight loss. Patients may have other signs of iron deficiency anemia like glossitis, angular cheilitis, koilonychias, splenomegaly may also be observed.[2]

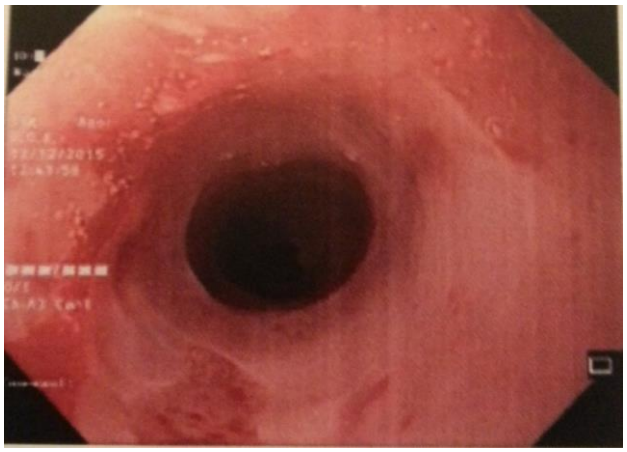

Figure 4: Endoscopic findings show web in oesophagus.

Iron deficiency anemia has been strongly associated with PVS as correction of anemia usually regresses the signs and symptoms of this disorder and also relieves dysphasia in some patients.[3] Genetic predisposition has been linked with PVS as multiple members of the family could be involved. Even autoimmune processes such as celiac disease, rheumatoid arthritis, thyroditis have been associated.[4] Diagnosis is established by confirmation of iron deficiency anemia with demonstration of web by upper GI endoscopy. Management involves correction of anemia, endoscopic dilatation, balloon dilatation, or incision of oesophageal web.[5] In view of risk for malignant changes in upper gastrointestinal tract, patients require regular follow-up and yearly endoscopic evaluation at least for initial few years after management.[6-8] Family history should be sought in such patients to rule out if any other family member was affected.

CONCLUSION

Plummer Vinson syndrome is a rear disease usually affecting middle aged females. It forms an important differential diagnosis of dysphagia. Since the dysphagia is slowly progressive, patients often present with iron deficiency anemia and treatment of anemia usually regresses signs and symptoms of PVS. Endoscopic dilatation or incision of esophageal web leads to marked symptomatic improvement in short span of time.

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