Plummer Vinson Syndrome in males: A rare case report

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Abstract

Plummer-Vinson syndrome (PVS) is characterized by dysphagia, iron deficiency anemia, and upper esophageal web. Exact data about epidemiology of the syndrome are not available: the syndrome is extremely rare. The associated symptoms can be resolved by administering iron supplements as well as by endoscopic intervention. Relapse in patients with Plummer-Vinson syndrome is very rare. There is an increased risk for squamous carcinoma of the pharynx and esophagus. We describe a case of a 47-year-old man with Plummer-Vinson syndrome whose symptoms were successfully treated with iron supplementation and blood transfusion at admission; however, investigations like endoscopy revealed esophageal web. This experience indicates that continuous iron supplementation and long-term follow-up is important in patients with Plummer-Vinson syndrome.

Key words: Plummer-Vinson syndrome, Squamous carcinoma, iron deficiency anemia, esophageal web

Introduction

Plummer-Vinson syndrome (PVS) is characterized by dysphagia, iron deficiency anemia and upper esophageal web. It is known by other eponyms as Paterson-Kelly syndrome, Paterson-Brown Kelly syndrome, Waldenstrom-Kjellberg syndrome, Sideropenic dysphagia[1]. It was first described in 1912 by Henry Stanley Plummer[2]. It is common in middle aged females, in fourth to seventh decade of life[3,4], with the ratio of female to male reported 4:1[3,4]. We report a case of this uncommon syndrome in a male.

Case Report

A male patient of age 47yrs presented with complains of easy fatigability, palpitation and exertional breathlessness since 6 months. On examination he was severely pale, with koilonychias (Figure 1), soft systolic murmur on cardiovascular examination, with the other systems being normal.

Investigations for cause of anemia revealed microcytic hypochromic anemia in peripheral smear with Hb 1.9g/dl with serum ferritin levels 3.47 ng/ml. On not able to find a definitive cause for iron deficiency anemia, history was revisited and the patient complained of mild dysphagia since 1yr for solids greater than liquids. Upper gastrointestinal endoscopy was done which showed: upper esophageal web (Figure 2) with the endoscope not able to passed through the web and was eventually ruptured. Blood transfusion and symptomatic treatment was given and patient symptoms improved. On follow up, repeat blood picture showed hemoglobin of 11 g/dl and patient is doing well with iron supplements.

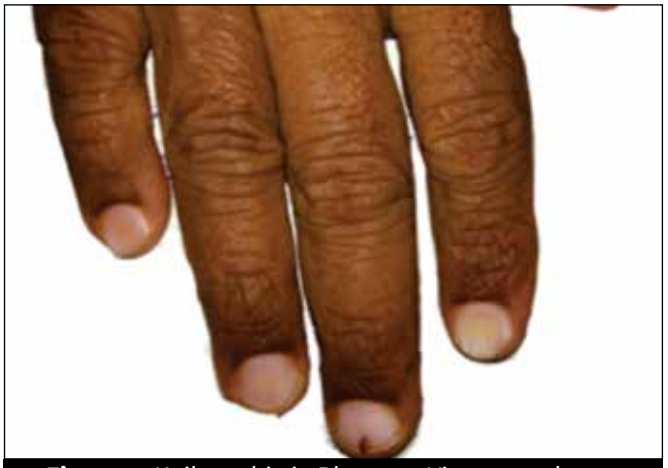

Figure 1. Koilonychia in Plummer-Vinson syndrome

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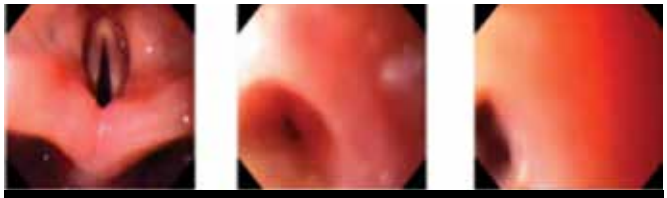

Figure 2. Upper esophageal web in Plummer-Vinson Syndrome

Discussion

Plummer-Vinson syndrome, also known as Paterson-Kelly syndrome or sideropenic dysphagia, is characterized by dysphagia, iron deficiency anemia, and mucosal lesion of the oral cavity or pharynx[5].

The syndrome is usually asymptomatic, but the patients sometimes develop dysphagia, weight loss, and weakness. Other symptoms may include nail deformation of the hand and foot, cheilosis, atrophic glossitis, early loss of teeth, conjunctivitis, dermatitis seborrhoea, hyperkeratosis, keratitis, blepharitis, visual disturbances and 30% of cases accompany splenomegaly[5]. The diagnosis of upper esophageal web is confirmed by radiologic methods or endoscopy. The radiologic method, however, is more suitable because endoscopy can sometimes miss the point of benign stricture, and does not verify most of the motility disorders[6].

Plummer-Vinson syndrome is known to be associated with an increased risk of upper alimentary tract cancers, and the incidence rate of upper esophageal cancer in this syndrome is 3–15%[7].

Management consisting of correction of iron deficiency in PVS may result in resolution of the associated dysphagia as well as disappearance of the web[8]. The esophageal web can be managed with endoscopic dilatation, balloon dilatation, endotracheal dilatation, and incision of web[2,4]. On occasion, more than one sitting may be required. Patients however require follow-up given the incidence of squamous cell carcinoma in up to 15% of the cases[9].

Conclusion

Esophageal web (Plummer Vinson syndrome) is usually seen in women and is rarely documented in males. A careful history taking and knowledge of occurrence of this rare disorder will benefit patients. It is an important syndrome because it identifies a group of patients at increased risk for squamous carcinoma of the pharynx and esophagus. Correction of iron deficiency in PVS may result in resolution of the associated dysphagia as well as disappearance of the web.

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