

Case Report

Nail: a window to systemic disease

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ABSTRACT

Plummer-Vinson syndrome is a rare clinical entity characterized by the triad of iron deficiency anemia, esophageal webs and dysphagia. The condition may manifest with the features like cheilitis, glossitis, atrophy of papilla over tongue, koilonychia indicating the underlying iron deficiency anemia and vitamin deficiency. We report a case of Plummer-Vinson syndrome presenting with koilonychia and cheilitis. Clinical, laboratory and endoscopic evaluation of the patient revealed diagnosis of Plummer-Vinson syndrome. Patient was advised iron supplementation which significantly improved patient's hemoglobin levels and with that the associated symptoms.

Keywords: Plummer-Vinson syndrome, Esophageal webs, Dysphagia, Koilonychia

INTRODUCTION

Nail abnormalities may be seen in a wide variety of systemic disorders. Though, most of these nail manifestations are non-specific, they should alert the dermatologist to the possibility of an underlying systemic disorder. These manifestations should be properly recognized and systematically evaluated for appropriate diagnosis and treatment. The importance of understanding these manifestations also lies in the fact that these nail symptoms and signs may be the first or the only clinical presentation.¹

Patterson-Kelly-Brown syndrome also known as Plummer-Vinson syndrome (PVS) is one such syndrome characterized by triad of chronic iron deficiency anemia, dysphagia and esophageal webs. This disease is usually seen in white women of 40–70 year age group.² Male population are less commonly affected. Thorough history taking, general clinical examination, hematological investigations and radiological examination are required for diagnosis of Plummer-Vinson syndrome. In this case

report we present a male patient, who presented with classical symptoms of PVS.

CASE REPORT

A 40-year-old male, came with abnormal nails and burning sensation of mouth since 2 years. On further inquiry he gave a history of dysphagia, easy fatigability. On general examination, patient's palpebral conjunctiva was pale, signifying anemia and the finger nails were spoon shaped i.e.; koilonychias (Figure 1). Intraoral examination revealed difficulty in mouth opening, angular stomatitis, erythema over the right buccal mucosa and loss of papilla over the dorsum of the anterior tongue (Figure 2). On further evaluation, hemoglobin was 6.8%, RBC-4.2 million/cumm, PCV-28%, MCV-60.5fl, MCH-15.3 pg, MCHC-25.4 gm/dl, RDW-18.1%, peripheral smear revealed hypochromasia with anisopoikilocytosis comprising of normocytes, microcytes and few elongated cells, indicating profound iron deficiency anemia. Subsequently, esophago-gastro-duodenoscopy report showed post-cricoid web. Hence a diagnosis of Plummer-Vinson syndrome (PVS) was made. Patient was given

oral iron supplementation and at 3 month follows up, patient shown significant improvement in hemoglobin levels.



Figure 1: Koilonychia (spoon shaped nails).



Figure 2: Loss of papillae over dorsum of the tongue.

DISCUSSION

Plummer first reported diffuse esophageal dilatation and upper esophageal spasm without anatomical stenosis, based on his previous work with 40 cases of cardiospasm.³ Subsequently a relation between the dysphagia and angulation of the esophagus with three characteristic manifestations: anemia, dysphagia, and atrophic glossitis was described by Porter Paisley Vinson in 1919.¹ Since then, the syndrome associated with these symptoms has been called Plummer Vinson syndrome. PVS is also termed as Paterson-Kelly syndrome and also known as Sideropenic dysphagia.

PVS is a very rare disease. Prevalence has been around <1/1,000,000. Female population in middle age is more often affected with a female to male ratio of 4:1. Recently, few reports of PVS in male patient are being

published leading to notion that PVS might be common in both males and females.⁴

The pathogenesis of the syndrome is unclear but iron deficiency anemia, malnutrition, genetic predisposition and autoimmune etiologies are postulated.^{5,6} As iron supplementation improves the dysphagia and esophageal webs, it is postulated that iron deficiency anemia is the widely accepted etiology. Iron-dependent oxidative enzymes get reduced due to iron deficiency which results in gradual degradation of the muscles of the pharynx leading onto mucosal atrophy and development of webs which causes neoplastic changes in the lower pharynx and upper esophagus.⁶ It is postulated that the high cellular turnover rate in the epithelium of the upper digestive tract makes it vulnerable to iron deficiency because of the deficiency of the iron- dependent enzymes. Dysphagia usually presents intermittently or progressively over years and is usually painless.

The other manifestations of Plummer Vinson syndrome include oral lesions like stomatitis, angular cheilitis, glossitis etc. and cutaneous manifestations namely koilonychia, dystrophy of nails. Koilonychia (spoon-shaped nails) are usually associated with chronic iron deficiency anemia occurring more commonly in adult females.⁷ Iron deficiency usually occurs secondary to gastrointestinal losses, nutritional deficiency, and/or malabsorption (e.g.,: celiac disease) or intestinal worms (e.g.,: Hookworm infestation).⁸ Women suffering from menorrhagia/ menometrorrhagia are often affected by iron deficiency. Iron supplementation not only improves the esophageal webs and dysphagia, it also improves the iron deficiency and with it the features of koilonychia and glossitis.

CONCLUSION

Nail abnormalities can be a revealing sign of a systemic disease may be a part of the puzzle for confirmation of a systemic disease. Koilonychia must prompt the dermatologist to actively search for the cause of iron deficiency and accurate diagnosis and appropriate treatment of the underlying condition is warranted. Plummer Vinson Syndrome is considered to be a premalignant condition and early diagnosis is of utmost importance for a better prognosis. As PVS is a precancerous condition with high malignant potential, early diagnosis is of utmost importance for better prognosis.

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