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Review Article

PLUMMER VINSON SYNDROME–A PREMALIGNANT CONDITION – AN OVERVIEW OF LITERATURE

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ABSTRACT

Plummer Vinson syndrome, (also known as Patterson Kelly brown syndrome, sideropenic dysphagia, hysterical dysphagia) is a premalignant condition, in which the patient has iron deficiency anemia, dysphagia and possibly a post-cricoid web that can predispose to malignant change. The oral mucosal changes such as atrophy, dryness, stomatitis and soreness may extend to the pharynx and upper part of the esophagus, causing spasm in the throat while swallowing. Diagnosis of the esophageal webs may be made by barium swallow, and then taking the radiographs. Usually, the dysphagic condition reverts back once the iron supplementation is instituted to the patient. However in some resistant cases, dysphagia donot respond to iron replenishment and require endoscopic dilatation. Hereby, presenting an overview of literature of plummer Vinson syndrome with a mention on the carcinomatous potential of this disorder.

Keywords: Plummer Vinson syndrome, Oral manifestations, esophageal webs, Postcricoid carcinoma.

INTRODUCTION

A wide range of hematological disorders encountered in internal medicine has their manifestations in the oral cavity and facial region[1]. Most of these manifestations are non specific, but should alert the hematologist and dental surgeon to the possibility of an underlying hematological disorder or a latent one that may subsequently manifest itself[1]. These manifestations must be properly recognized if the patient is to receive appropriate diagnosis and referral for treatment. The importance of understanding orofacial manifestations of these disorders also lies in the fact that orofacial signs and symptoms may be the first clinical presentation that may alert the dentist / hematologist to an underlying hematological disorder. Disorders of the erythroid (anemia and polycythemia), lymphoid and megakaryocytic-platelet compartment of the bone marrow, immune system deficiencies, coagulation as well as human immunodeficiency virus infection have been widely reported to manifest in the or facial region[1,2,3].

Plummer-Vinson syndrome is a rare syndrome characterized by dysphagia, esophageal webs and chronic iron deficiency anemia. Oral characteristics include glossitis, glossopyrosis, glossodynia, and angular cheilitis[4]. Its etiology is unknown although autoimmune, genetic, infectious and nutritional factors have been proposed as a cause[5]. Approximately 10% of patients suffering Plummer-Vinson syndrome develop squamous cell carcinoma principally in the hypo pharynx and esophagus[6]. It is also associated with koilonychias (spoon shaped nails), pagophagia and dysphagia due to pharyngoesophageal ulcerations and esophageal webs[7].

DISCUSSION

In 1912, Plummer[8] reported that there were some cases of dysphagia associated with severe anemia which had been regarded as a kind of hysteria or neurosis of unknown cause. Subsequently, Vinson[9] reported that this type of dysphagia has three characteristic manifestations: anemia, dysphagia and atropic glossitis, and Kelly[10] and Paterson[11] pointed out the high incidence of hypo chromic anemia in this disease. Since then, the syndrome associated with these symptoms has been called Plummer-Vinson syndrome or Paterson-Kelly syndrome. This syndrome is characterized by three main symptoms-atrophic changes in the mucosa of the oral cavity, pharynx and esophagus, dysphagia and hypochromic anemia. Other oral symptoms are associated such as angular cheilosis, early loss of teeth, glossitis, etc., as well as other various signs such as esophagism, cardiospasm, web formation, achlorhydria, nail deformation, splenic tumors, dermatitis seborrheica, hyperkeratosis, conjunctivitis, keratitis, blepharitis, and visual disturbances, which may also occur as complications. Rare manifestations such as clubbing instead of

koilonychia and tortuous esophagus in addition esophageal webs have also been described[12]. Symptoms secondary to anemia such as pallor, fatigue and weakness may also dominate the clinical picture.

The syndrome mainly affects white women, in the 4th to 7th decade of life, but some pediatric and adolescent cases have also been reported[4,13,14,15]. The disease is rarer nowadays and the incidence is decreasing even in African nations where iron deficiency and malnutrition are common[16]. The incidence and prevalence are not reliable as hematological parameters are not included[17].

Plummer Vinson syndrome is a manifestation of severe, long term, iron deficiency anemia causing dysphagia because of esophageal webs. While esophageal webs are thin mucosal folds (covered by squamous epithelium) protruding into the lumen of proximal esophagus, esophageal rings occur more distally and are further classified as muscular A rings (upper part of distal esophagus), mucosal Schatzki B rings, and distal most C rings[18]. Dysphagia from the webs is usually painless and intermittent or progressive.

The pathogenesis of the syndrome is not clear, but iron deficiency anemia, malnutrition, genetic predisposition, autoimmune etiologies are postulated[4,19]. Iron deficiency anemia is most widely accepted etiology as dysphagia and esophageal webs improve with iron supplementation[4]. Upper esophageal webs are seen in only 10% of patients with iron deficiency which denotes multiple etiologies in this syndrome[20]. Iron deficiency causes reduction of iron dependent oxidative enzymes which results in gradual reduction of the muscles of pharynx leading onto mucosal atrophy and esophageal webs[19].

Most commonly, patient first have dysphagia to solids, but over time, symptoms can progress to involve dysphagia to liquids as well[14]. According to Novacek[4] the dysphagia is usually non painful, and it's progression can eventually lead to weight loss[7,21]. Logan [22] reported that patients often tolerate progressive dysphagia for considerable period of time without seeking medical attention, thus leading to late presentation.

Patients with Plummer Vinson syndrome usually complain of dysphagia and anemic symptoms and first visit otorhinolaryngologists or internists. Regarding dysphagia, many reports guery whether it precedes or follows anemia, and whether its cause is a functional or organic disturbance. At first, hysteria or spasm of the upper esophagus was thought to cause dysphagia and then anemia and nutritional disorder were thought to be triggered[8,9]. Subsequently, it was thought that iron deficiency or anemia leads to mucosal atrophy in the digestive tract, ulcerous changes or cracks may occur at the inlet of the esophagus as trauma from the intake of solid foods, and then a web is induced, which results in organic stricture, and this may finally cause dysphagia[23,24].

PVS has been identified as a risk factor for developing squamous cell carcinoma of the upper gastrointestinal tract. Three to fifteen percent of the patients with PVS, mostly women between 15-50 years of age, have been reported to develop esophageal or pharyngeal cancer[25,26]. According to Kim *et al* [27], PVS is thought to be precancerous because squamous cell carcinoma of hypo pharynx, oral cavity or esophagus takes place in 10% of those patient suffering from this malady. The incidence of cancer of the tongue while comprising 25-50% of all intra-oral cancers is relatively uncommon among women except in certain geographical localities chiefly Scandinavian countries, where it has been been associated with pre-existing PVS[28,29].

However, according to Uygur-Bayramicli *et al*[29]; some patient with all of the classical features of PVS will present with esophageal stricture instead of web. A barium swallow study is advised for the detection of a web, and other method will be an upper gastrointestinal endoscopy[4,20].

The treatment of PVS is hinged on the clarification of the cause of iron deficiency if a patient demonstrate one, and the rupture of and dilation of the web if it causes obstruction[4,30,31]. The prognosis of this condition is good as anemia and dysphagia can be effectively treated by iron therapy and webs by dilatation[32]. The prognosis worsens dramatically if the syndrome is associated with complications like squamous cell carcinoma of hypo pharynx and upper esophagus[4]. Endoscopic dilatation is the procedure of choice for treating esophjageal webs.

CONCLUSION

Mouth is the mirror of systemic diseases. Many systemic diseases may be diagnosed on the basis of their oral manifestations. Plummer Vinson syndrome is a form of iron deficiency anemia, in which the patient has signs of iron deficiency (pallor, fatigue, oral dryness, depopulation, burning sensations), dysphagia and esophageal webs. The condition is precancerous with high potential of developing into post-cricoid carcinoma. Early diagnosis is essential for appropriate treatment of the condition.

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