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Case Report

Oral Manifestations of Plummer-Vinson Syndrome: A Classic Report with Literature Review

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Abstract:

Plummer-Vinson syndrome (PVS) is a triad of microcytic hypochromic anemia, atrophic glossitis, and esophageal webs or strictures. It is one of the syndromes associated with iron deficiency anemia. Symptoms resulting from anemia predominates the clinical picture, apart from the additional features such as glossitis, angular cheilitis, and dysphagia. Dysphagia is main clinical feature of PVS. PVS carries an increased risk of development of squamous cell carcinoma of esophagus and pharynx. A classic case report of PVS with clinical features, oral manifestations, malignant potential, differential diagnosis, investigation, dental implication, and treatment is discussed here with the literature review from the dentist's point of view. The article carries a message that dental surgeons have to be familiar with the oral manifestations of anemia and be able to suspect PVS to aid in early diagnosis and prompt treatment.

Key Words: Dysphagia, iron deficiency anemia, Plummer-Vinson syndrome

Introduction

Plummer-Vinson syndrome (PVS) is known by several names such as Paterson-Kelly syndrome, Paterson-Brown Kelly syndrome and sideropenic dysphagia. PVS was first described by Paterson and Kelly in 1919. It is named after Henry Stanley Plummer (1874-1936) and Porter Paisley Vinson (1890-1959) who were physicians of the Mayo Clinic. Donald Ross Paterson (1863-1939) and Adam Brown-Kelly (1865-1941), both British laryngologists published their findings independently in 1919.[1]

PVS has become increasingly rare with the availability of iron supplements, but nonetheless it should be suspected in cases of iron deficiency and dysphagia. The syndrome consists of glossitis, atrophic oral mucosa, dysphagia, and anemia. Post-menopausal women are commonly affected. Other presenting symptoms may include fissuring at the angles of the mouth and glossodynia. PVS is a potentially malignant disorder with the risk of squamous cell carcinoma of the esophagus due to the post cricoidal webs that are formed.[2] This article presents a classical case of PVS with review of literature with dental implications.

Case Report

A 20-year-old female reported to the dental outpatient department with a complaint of difficulty in swallowing for the past 6 months. Her medical history was non-contributory. On eliciting her personal history, it was found that she had increased blood loss during menstruation.

On examination, she appeared to be poorly built and moderately nourished. Extraoral examination revealed pallor of the palpebral conjunctiva (Figure 1a) and koilonychia in all her fingers (Figure 1b).

Intra oral examination showed the presence of ulcers at the angle of mouth with pigmented spots on the tongue (Figure 2a) she had a restricted mouth opening of 25 mm. Mucosal examination revealed pallor of the labial, buccal, palatal mucosa, retromolar area, and tongue. Marked mucosal pigmentation was observed in the buccal and labial mucosa (Figure 2b).

Hematological investigations revealed her hemoglobin count to be only 4 mg/dl, examination of the peripheral blood smear showed the distinctive picture of microcytic hypochromic anemia (Figure 3).

Radiograph examination consisted of barium swallow test which revealed the constriction of esophagus (Figure 4a). Endoscopy confirmed the presence of esophageal web (post cricoids web 17-18 cm) and also a ruptured web (Figure 4b). The instrument did not pass through at the level of the web. On correlating the clinical history, examination and investigations performed a final diagnosis of PVS was concluded.

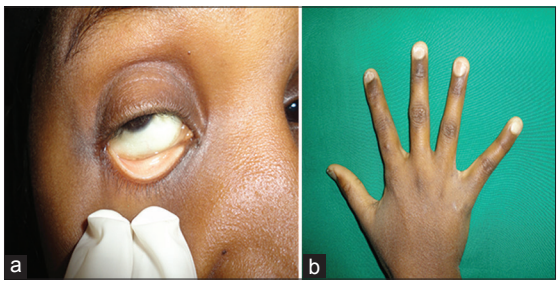

Figure 1: (a) Extra oral view showing a pale conjunctiva. (b) Koilonychia evident in nails.

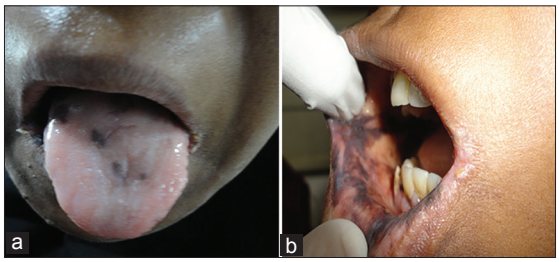

Figure 2: (a) Atrophy of tongue and angular cheilitis. (b) Marked intraoral pigmentation.

The patient is currently managed by iron supplementation in the form a loading dose of Fe dextran give intramuscular with an oral maintenance dose of ferrous sulfate 325 mg (60 mg of iron) given thrice daily. Mechanical dilation of the post cricoidal webs through endoscopy is being carried out on a regular basis, and the patient has reported reduction in dysphagia.

She is under constant follow-up and review along with a well-maintained high vitamin supplemented diet since the post cricoidal webs have a premalignant potential.

Discussion

Anemia is a disordered process in which the rate of red cell production fails to match the rate of destruction, which results in a reduction of hemoglobin concentration. The etiopathogenesis of anemia revolves around increased physiological demand, pathological blood loss and inadequate iron intake.[3] Food sources rich in iron are the main source, and iron is recycled by the destruction of senescent red blood cells. Iron is important in the body for maturation and to maintain the integrity of the epithelium.

Iron deficiency anemia is caused mainly by the lack of iron and anemia develops when there is inadequate iron for hemoglobin synthesis. Peripheral causes include blood loss, hemolysis, and hypersplenism. Females in reproductive age group, menstruation, pregnancy, nutritional deficiency, blood loss, and malabsorption are all known predisposing factors.

Etiopathogenesis

Etiopathogenesis of PVS is uncertain. But iron deficiency, malnutrition, genetic predisposition or autoimmune processes

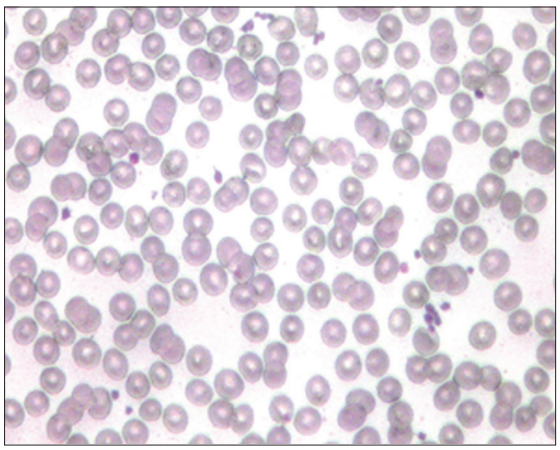

Figure 3: Peripheral blood smear showing microcytic hypochromic anemia (Leishman's Stain at ×100 magnification).

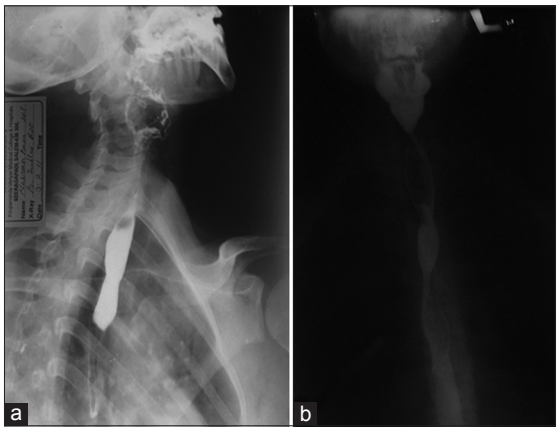

Figure 4: (a) Barium swallow – lateral view. (b) Barium swallow showing esophageal web.

may be the contributory factors of this syndrome. Myasthenic changes occur in muscles involved in the swallowing mechanism due to the depletion of iron-dependent oxidative enzymes. Atrophy of the esophageal mucosa and formation of webs is seen as mucosal complications. These changes were evident in the present case.

An autoimmune mechanism is certainly associated as the syndrome is seen in association with rheumatoid arthritis, thyroiditis, celiac disease, and pernicious anemia. Other factors such as nutritional deficiencies, genetic predisposition are thought to play roles in the causation of this disease.

Clinical features

The main clinical features of PVS are dysphagia, iron deficiency anemia with upper esophageal webs. Majority of the patients are women in the fourth to sixth decade of life, the syndrome has also been described in adolescents and children.

The dysphagia usually presents intermittently or progressively over the years and is usually painless. It is also characterized by enlargement of the spleen and thyroid.

Epithelial changes include koilonychia or spoon shaped nails, atrophic glossitis, angular stomatitis, dryness, and atrophic changes in the conjunctiva along with the formation of post cricoidal esophageal webs. All these classic findings were found in the present case.

Oral manifestations include stomatitis, angular cheilitis, glossitis with different degree of atrophy of fungiform and filiform papilla, recurrent aphthous stomatitis, pale oral mucosa, oral candidiasis, erythematous mucositis, and a burning mouth. The atrophy of filiform papillae are most susceptible to nutritional deficiency and disappear first, followed by fungiform papillae, regeneration occurs in reverse order but vallate and foliate papillae on the posterior third are spared.

Diagnosis

The diagnosis of iron deficiency anemia relies on clinical history with questions about dietary intake, and the presence of blood in stools (which may be a sign of hemorrhoids. In women, a careful inquiry about the duration of periods.3 Clinically, the patient will present with pallor.

Hematological investigations revealed an iron deficiency anemia with decreased values of hematocrit, hemoglobin, serum iron, mean corpuscular volume, and increased total iron binding capacity. A pathognomonic picture of microcytic hypochromic anemia is seen. A similar picture was seen in the present case.

In PVS, radiographic examination of the pharynx will show the presence of esophageal webs2 along with other manifestations like koilonychia.

Differential diagnosis

It includes all the possible causes of dysphagia like malignant tumors, benign strictures, diverticula, achalasia, spastic motility disorders, scleroderma, gastroesophageal reflux disease, neuromuscular and skeletal muscle disorders.4

Malignant potential

PVS is associated with a high risk of development of squamous cell carcinoma of esophagus and pharynx. A post cricoid web is a rare complication of iron deficiency anemia.

The mechanism explained is that the anemia causes epithelial atrophy, changes in cell kinetics, and decreases the repair capacity of the mucosa. This allows the carcinogens and cocarcinogens to act aggressively, predisposing the entire oral cavity and esophageal area to malignancy.[5]

PVS is a major risk factor for the development of squamous cell carcinoma of the upper gastrointestinal tract, about 3-15% of the patients with PVS, has presented with esophageal or pharyngeal cancer.[6]

Implications of anemia in dentistry

1. Mucosal disease
   - Glossitis
   - Angular stomatitis
   - Recurrent aphthae.
2. Infections like candidiasis
3. Risks from general anesthesia
   - Shortage of oxygen can be dangerous.

Management

Correction of etiology is the mainstay of treatment. Iron supplementation in the form of ferrous sulfate orally can be given or parenteral doses of iron in severe cases. Iron supplementation alone can resolve dysphagia in many patients.

Mechanical dilation of webs and strictures are done by endoscopic dilatation in single or multiple sessions depending on the severity of the web, in which wire-guided bougies or balloons are used to disrupt and rupture the fibrous tissue of the stricture.7

For tight or highly constricted webs which prevent the passage of the endoscope a fluoroscopic guidance is usually required. Endoscopic balloon dilation becomes difficult when the webs are located proximally, but the same can be performed under fluoroscopic guidance.[8]

Other successful means of disrupting an esophageal web are by ND:YAG laser therapy or needle-knife electro incision which is seldom used.

Patient should be kept on a high vitamin diet to maintain integrity and maturative potential of the oral epithelium since PVS is a potentially malignant disorder. Regular follow-up is mandatory. It is wise to perform an upper gastrointestinal endoscopy annually.

Conclusion

Iron deficiency is a common cause of anemia. Severe iron deficiency anemia with koilonychia and dysphagia are the features of PVS. PVS is considered to be a premalignant condition, and dental surgeons have to be familiar with the oral manifestations of anemia and should be able to suspect PVS. Early diagnosis is of utmost importance for a better prognosis. A case of PVS with barium swallow performed has been described here with the literature review.

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