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Oral Health - A Mirror of Plummer Vinson Syndrome

Dr. Saraswathi Gopal. K[1], Dr. Mahesh Kumar. P[2], Dr. Alice.N[3]

[1]Professor and Head of the Department, Department of Oral Medicine and Radiology, Faculty of Dentistry, Meenakshi Ammal Dental College and Hospital, Meenakshi Ammal Higher Education & Research Institute, Chennai-95
[2]Associate Professor, Department of Oral Medicine and Radiology, Faculty of Dentistry, Meenakshi Ammal Dental College and Hospital, Meenakshi Ammal Higher Education & Research Institute, Chennai-95
[3]Post Graduate, Department of Oral Medicine and Radiology, Faculty of Dentistry, Meenakshi Ammal Dental College and Hospital, Meenakshi Ammal Higher Education & Research Institute, Chennai-95

Corresponding Author: Dr. Alice Josephine Rani. N

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ABSTRACT

A lack of significant iron in the normal red blood cells leads to iron deficiency anaemia (IDA). Plummer Vinson Syndrome (PVS) is a syndrome with the clinical triad of symptoms of iron deficiency anaemia, dysphagia and oesophageal webs. Repeated injury to epithelia due to iron deficiency leads to atrophy of mucosa and degradation of pharyngeal muscles, leading to the development of oesophageal webs. Oral manifestations are the initial symptoms and oral cavity is the mirror of the systemic conditions of the body. Henceforth dentists play a major role in promptly diagnosing the oral health of an individual. Under this background, we present a case of Plummer Vinson Syndrome with oral manifestations.

Key words: Anaemia, Iron deficiency, Plummer Vinson Syndrome, Haemoglobin, Dysphagia, Oesophageal webs, Endoscopy, Oral Health.

Abbreviations: IDA – Iron Deficiency Anaemia; PVS – Plummer Vinson Syndrome; H/O – History of; WHO – World Health Organisation

INTRODUCTION

Blood constitutes 7-8% of human body weight. This fluid carries out the critical functions of transporting oxygen and nutrients to our cells and getting rid of carbon dioxide, ammonia, and other waste products. Additionally, it plays a vital role in our immune system and in maintaining a relatively constant body temperature. Blood is a highly specialized tissue composed of more than 4,000 different kinds of components. Red cells, white cells, platelets, and plasma are the four most important cells[1]. Iron is a vital element for blood production. About 70 percent of your body's iron is found in the red blood cells called haemoglobin and in muscle cells called myoglobin. Haemoglobin carries oxygen in blood from the lungs to the tissues. Myoglobin accepts, stores, transports and releases oxygen[2]. A lack of significant iron in the normal red blood cells leads to iron deficiency anaemia (IDA). It is the most common type of anaemia especially in children and women. The three well known stages of iron deficiency anaemia are pre-latent, latent and the marked iron deficiency[3]. Plummer Vinson Syndrome (PVS) is a clinical triad of symptoms of Iron Deficiency anaemia, dysphagia and oesophageal webs. Difficulty in swallowing is a common symptom and oesophageal webs are an infrequent cause of dysphagia. It has been named variously as "Plummer–Vinson syndrome" (PVS;

primarily in the USA) and as "Paterson–Brown–Kelly syndrome" (used primarily in the UK)[1]. The former term was derived from the names of two physicians – Henry Stanley Plummer in 1912 AD and Porter Paisley Vinson in 1919 AD, at the Mayo Clinic – who separately described this condition[4].

The exact pathogenesis of PVS and the formation of the oesophageal web is unknown. It has been postulated that iron deficiency induces iron-dependent enzyme dysfunction, leading to oxidative stress and DNA damages in the epithelia of the oesophageal mucosa[5]. Repeated injury to epithelia due to iron deficiency leads to atrophy of mucosa and degradation of pharyngeal muscles, leading to the development of oesophageal webs. The oesophageal web is localized below the cricopharyngeal muscle and is asymmetrically attached to the anterior oesophageal wall. The oesophageal web is a thin mucous membrane, composed of squamous epithelia[6].

CASE REPORT

Here we present a case report of Plummer Vinson Syndrome in a 25 years old young female patient with oral manifestations.

A 25 years old female patient came with the chief complaint of pain in the left lower back tooth region for the past 10 days. Patient gave history of an intermittent and dull pain which aggravates on eating and relieves by itself. On Past Medical History, Patient gave H/O symptoms of anaemia 5 years before for which she was hospitalized for 15 days and she felt better until before 3 months. For the past 3 months patient had symptoms of burning sensation of mouth, difficulty in swallowing and consulted a physician before 10 days and advised to do blood tests for anaemia. The recent report reveals haemoglobin 4.5mg/dl and Ferritin levels with 9.2ng/ml and she was advised for blood transfusion. Patient also gave H/O drug allergy for pain killers which manifested as ulcers and mild swelling of face. On General examination, Patient was poorly built and poorly nourished. Her lemon-tinted skin, clubbing nail beds, loss of appetite, difficulty in swallowing and stomach burning sensation were present. On extra oral examination, presence of paleness of right & left lower palpebra (Figure – 1). Lips were pale, dry with evidence of encrustations on the right & left commissures. On Intra oral examination, presence of pale mucosa with diffuse brownish pigmentation from the commissures of the lip on both the sides and depapillation of tongue with a glossy and bald appearance (Figure – 2), Dental Caries: 14, 17, 25, 28 & 37, Grossly decayed: 36, Root Stumps: 23, 47. Moderate amount of plaque and calculus was present. Correlating the clinical findings, patient's history and basic blood investigations, a provisional diagnosis of Plummer Vinson syndrome was given. Further patient was subjected to peripheral blood smear and endoscopy. Peripheral blood smear revealed RBC's appearing as microcytic hypochromic cells with moderate anisocytosis, tear drop and pencil cells. Endoscopy (Figure - 3) findings of Esophagus were mild constriction noted at the proximal aspect and laxity of lower esophageal sphincter; Duodenum appeared to be normal. All these investigation reports conclude a final diagnosis of Plummer Vinson Syndrome. Patient was referred to general physician for further management. Further patient was advised for pharmacological management, Iron supplements as well as transfusion of packed cell volume. After transfusion of 2 packed cell volume with Iron supplements and non-pharmacological management, patient had hemoglobin level of 8.8gm/dl within 45 days. Also, Parenteral administration of Iron injections was advised for 4 weeks by the Physician.

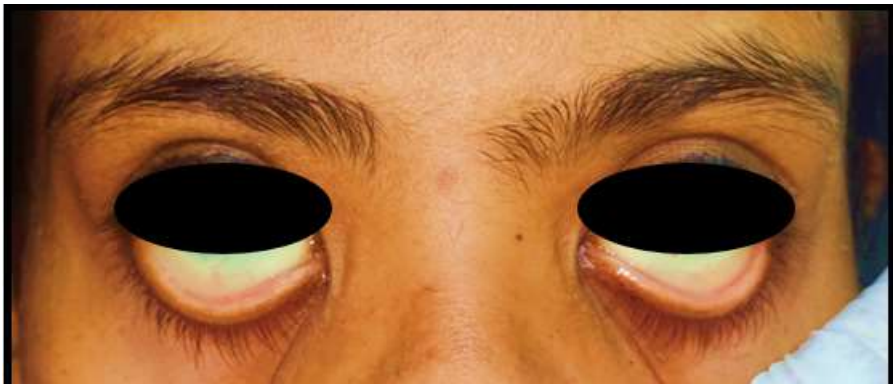

FIGURE – 1

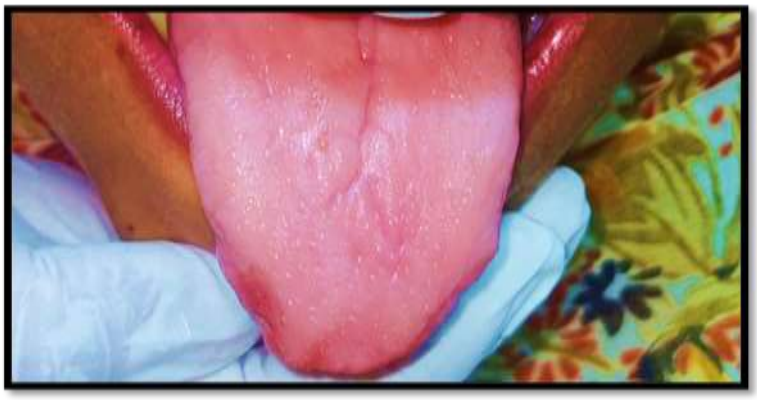

FIGURE – 2

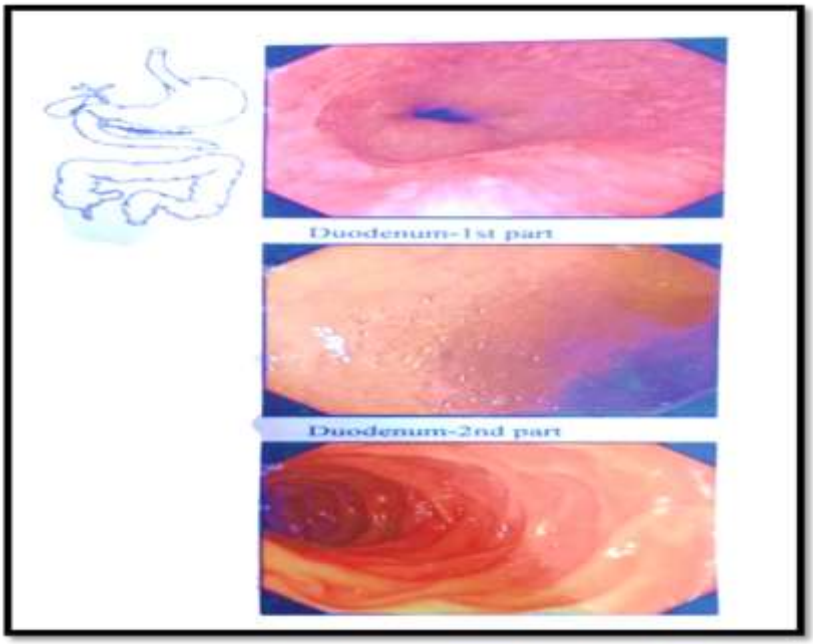

FIGURE – 3.

DISCUSSION

Anaemia is defined as reduction in the oxygen carrying capacity of blood, due to abnormality in Hb within Red blood cells or reduction in number of circulating RBCs. The World Health Organization (WHO) defines anemia as Hb< 13g/dl in men above 15 years, and < 12g/dl in women above 15 years of age[6]. Identifying anaemia is highly essential as it is a weakening condition that can induce or worsen an existing disease like congestive heart failure and can cause

cognitive impairment, dizziness and apathy impairing quality of life[3]. Causes of iron-deficiency vary with age, sex and country of residence of patient. In general, the causes of iron deficiency anemia can be grouped as: 1. Dietary inadequacy of iron as in milk fed infants, poor economic status individuals, anorexia; 2. Increased iron loss from the body, e.g. uterine bleeding in females in the form of excessive menstruation, repeated miscarriages, post-menopausal bleeding, gastrointestinal bleeding due to peptic ulcer, haemorrhoids, ulcerative colitis, renal tract bleeding, nasal bleeding, hemoptysis; 3. High demand of iron as in Infancy, childhood and adolescence, menstruating females and pregnant females. 4. Reduced iron absorption, as seen in partial or total gastrectomy, Achlorhydria and intestinal malabsorption diseases[7].

Haemoglobin is the most abundant iron containing protein in the human body which accounts for nearly one half of the total iron in the body. Tissue oxygenation, erythrocyte turnover, and erythrocyte loss from haemorrhage are the three variables that constitute Erythropoiesis-related demands of iron. Approximately 20 ml of senescent erythrocytes gets eliminated and among that 20mg of Iron is recycled for the production of new erythrocytes every day. In Iron Deficiency Anaemia, half-life of circulating Iron is shorter. Additional iron must be absorbed from the diet to meet the steady-state demands of the host suffering with any sorts of haemorrhage. When there is a continuous loss of iron, the newly produced erythrocytes will have decreased haemoglobin. The bone marrow tries to pump out all the RBC's before maturing to meet the demand, thus resulting in altered shape –Poikilocytosis and altered size – Anisocytosis of the cells[3]. The Grading system of dysphagia is as follows: grade I - occasional dysphagia on taking solids, grade II - able to swallow only semi-solid diet[6]. The dysphagia in PVS has a gradual onset, and is usually painless and intermittent. The site of obstruction is usually pointed to the neck at or above the suprasternal notch by the patients. It is noticed first for solid foods, which is then followed after several years by difficulty in swallowing liquids, suggesting an extremely slow progression of the oesophageal obstruction. The patients tend to get relieved of the symptom by either modifying the diet or consuming softer solids, and/or by enhanced chewing to break the food into smaller particles. This takes a longer time for completing a meal due to obstruction. Choking and/or episodes of aspiration may occur with severe obstruction[4]. Other clinical findings can be glossitis and angular cheilitis. Nutritional deficiencies may also get associated with Iron Deficiency Anaemia and their manifestations could be soreness of mouth, angular cheilitis, atrophic glossitis, koilonychia (spoon-shaped finger nails), clubbing, seborrheic dermatitis, hyperkeratosis, conjunctivitis, keratitis, paraesthesia and/or night blindness[8]. Occasionally, women may experience complaints of burning and itching in the vulval region and gets disappeared after iron treatment, which states that involvement of genital mucosa with epithelial lesions are similar to those in the mouth and throat[9].

The main objectives of investigations in a patient with suspected oesophageal web or Plummer Vinson Syndrome are: to diagnose anaemia, ascertain the cause of anaemia, assess the severity and cause of dysphagia and to localize the obstructing lesion to help plan definitive treatment[10]. Hence, the investigations include hematologic tests, and radiographic (barium swallow) and endoscopic examinations of the oesophagus[4].

Haemoglobin concentration in blood, microscopic examination of peripheral blood smear, red cell indices (mean corpuscular volume, mean corpuscular haemoglobin and mean corpuscular haemoglobin concentration) and serum iron studies (serum iron, ferritin and total iron binding capacity) are helpful in diagnosing the presence or absence of anaemia and to establish iron deficiency as its cause[4].

Barium swallow radiography is the investigation most commonly asked for if an oesophageal web is suspected. It helps to differentiate between benign and malignant causes of obstruction, planning of definitive treatment, and provides a reproducible documentation of pre-treatment status for comparison after treatment or other later use[9]. Fibre-optic endoscopy is a safe and reliable tool for examining gastrointestinal tract. Endoscopic examination of the oesophagus is called esophagoscopy. Esophagoscopy has the advantage of permitting treatment in the same sitting[4].

The management of these conditions are done through blood transfusions especially in the cases of acute blood loss. An oral iron therapy of 200 mg is generally prescribed for 6- 12 months where it is said that only 50 mg per day of iron absorption takes place. In cases where patient is intolerable to oral iron therapy a parenteral route of iron therapy is prescribed. The main complication in this route is the anaphylaxis[3].

CONCLUSION

Oral cavity is the mirror of the body and oral manifestations are the initial symptoms of systemic conditions of an individual especially anaemia. Dentists play a vital role to scrutinize in ruling out a prompt diagnosis of the oral health of an individual.

Conflict of Interest: None

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