

Plummer–Vinson Syndrome: A Case Report and Medical Management

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Plummer–Vinson syndrome (PVS) is a condition that can occur in people with long-term iron-deficiency anemia. It is a triad of dysphagia, iron deficiency anemia, and upper esophageal web(s). The post-cricoid web is associated with high-risk of development of squamous cell carcinoma of the esophageal region. Predominantly affected are middle-aged women in developing countries. Oral manifestations play an important part of the diagnosis of PVS. Timely diagnosis and treatment of this syndrome is important. An interesting case of PVS with typical clinical, oral and radiological findings previous to and post management of the syndrome are discussed in this report. It also includes a review of literature.

Keywords: Barium esophagogram, Burning sensation, Dysphagia, Iron-deficiency anemia, Post-cricoid web

INTRODUCTION

Patterson and Kelly first reported Plummer–Vinson Syndrome (PVS) in 1919.¹ The syndrome consists of a triad of dysphagia, iron deficiency anemia, and upper esophageal web(s). At the same time, it also consists of atrophic oral mucosa, cracks or fissuring at the corners of the mouth, glossitis, koilonychia, or nails that are brittle and break easily. Over years, dysphagia progresses, gradually, and the patient increasingly finds it difficult to swallow solid food. Middle-aged women are commonly affected, and it is very rare in childhood.² This syndrome is both of diagnostic and of prognostic significance.³ This case report presents a classic example of PVS, its dental implications and includes a review of the literature.

CASE REPORT

We report a case occurring in a 42-year-old woman. She presented with burning sensation in her mouth and progressive dysphagia especially in swallowing large morsels of food for the last 6 months. She had poor nutritional status. She is a known case of hypothyroidism

for the past 10 months and has been under medication. She complained of irregular menstrual cycles. She did not complain of loss of appetite or weight. There was no history of fever, nausea, vomiting, altered bowel habits, melena, or hematochezia. No history of ingestion of nonsteroidal anti-inflammatory drugs or any other drugs. History of alkaline or acid ingestion was negative.

The weight and height were within normal limits for her age and sex. Physical examination disclosed that her vital signs were normal, and no abnormality detected except marked pallor. In the lower palpebral conjunctiva and oral mucosa, pallor was evident. Angular cheilitis and glossitis were evident (Figure 1). Mouth opening was restricted (interincisal distance -16 mm). Auscultation of heart revealed no murmurs. Per abdomen, examination revealed it to be soft and nontender with no organomegaly.

Her laboratory data revealed iron deficiency anemia with a hemoglobin level of 7.4 g/dl, mean cell volume (MCV) 67.5 FL, mean cell hemoglobin (MCH) 19.7 pg, mean corpuscular hemoglobin concentration (MCHC) 28.7g/dl. Serum iron was 9 µg/dl. Peripheral smear revealed microcytic hypochromic cells. Occult blood in stools was present. Serum ferritin and total iron binding capacity was desired but could not be done as the tests were expensive and the patient could not afford them.

Radiological examination by barium swallow under fluoroscopic control showed the presence of an anterior

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Figure 1: Clinical appearance of (a) pallor in the lower palpebral conjunctivae, (b) glossitis, and (c) angular cheilitis

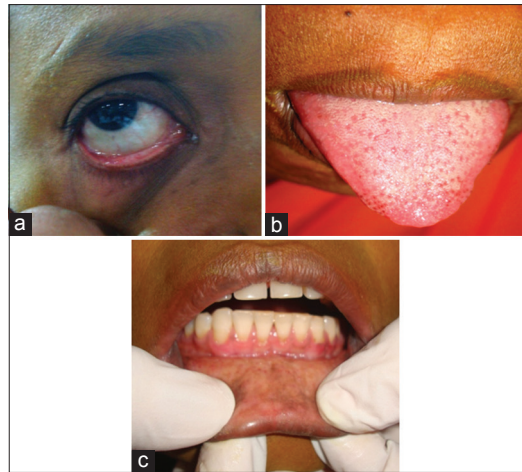


Figure 3: (a-c) Clinical appearance after treatment



Figure 2: Barium esophagram showed the presence of an anterior web at the level of cervical oesophagus at the C4 level

web at the level of the cervical esophagus at the C4 level (Figure 2).

The patient was hospitalized and treated with iron supplementation. At the end of 1 week of iron therapy and transfusion of two units of packed red cells, her fatigue and dysphagia reduced. Hemoglobin and MCV level divulged an improvement to 10g/dl and 72 fl , respectively.

She has been on regular follow-up ever since. She remains in good general condition without any complaint of dysphagia after 6 months of treatment. Mouth opening increased to 25 mm, burning sensation and angular cheilitis were relieved (Figure 3). She continued intake of 322 mg ferrous fumarate every day for the past 6 months. Follow-up of the patient with investigations after 6 months revealed hemoglobin level of 12.6 g/dl. Her MCV was 85.7 fl, mean cell hemoglobin 29.4 pg, MCHC 34.3 g/dl, serum iron 105 µg/dl. Peripheral smear revealed normocytic normochromic cells. Radiological examination by barium



Figure 4: Barium esophagram showed the presence of a small thin anterior shelf like filling defect seen at the C4 level which is decreased in size when compared to the previous study

swallow showed the presence of a small thin anterior shelf like filling defect seen at the C4 level (Figure 4), which is decreased in size when compared to the previous study.

DISCUSSION

The predominant clinical features of PVS are upper esophageal web(s), dysphagia, and iron deficiency anemia.¹ Most of the patients are middle-aged women, in the fourth to the seventh decade of life² similar to our patient. The dysphagia is progressive over years, limited to solids and

sometimes associated with weight loss. Anemic symptoms such as weakness, pallor, fatigue, and tachycardia may dominate the clinical picture. Furthermore, it is characterized by glossitis, angular cheilitis, and koilonychia. Enlargement of spleen and thyroid is also observed.¹ In the present case, the patient suffered from dysphagia and burning sensation in her mouth.

Esophageal webs can be detected by barium swallow X-ray. Barium sulfate is an inert material that produces good contrast. For better visualization, it is suggested that a thick paste should be used and rapid exposure after swallowing is essential,³ as done in our case. Presently, women have a higher incidence of PVS which is usually attributed to inadequate dietary intake and chronic blood loss in the form of menstruation.⁴

The etiopathogenesis of anemia is due to increased physiological demand, pathological blood loss, and inadequate iron intake.⁵ The theory of iron-deficiency anemia bases on the rapid losses of iron-dependent enzymes due to its high cell turnover. This reduction may cause mucosal degenerations, atrophic changes, web formation, and even lead to cancer development of the upper gastrointestinal tract.⁶ The depletion of iron-dependent oxidative enzymes may produce myasthenic changes in muscles involved in the swallowing mechanism, atrophy of the esophageal mucosa and formation of webs as epithelial complications.³ As anemia causes epithelial atrophy and decreases the repair capacity of the mucosa, it allows the carcinogens and cocarcinogens to act aggressively and predisposes the entire oral cavity and esophageal area to malignancy.³

In patients with hypothyroidism, the prevalence of anemia is shown to be 20-60%. This is based on the theory that thyroid hormones are involved in hemoglobin synthesis in adults and maturation of hemoglobin in the fetus. By affecting the hematopoietic process, hypothyroidism results in anemia through slowing the oxygen metabolism.³ This is in concordance with the existing case where the patient is a known case of hypothyroidism and is diagnosed with iron deficiency anemia.

As per a study done by Noshier *et al.*, in 1975, a series of 1000 consecutive patients underwent a cineradiographic examination of the hypopharynx and cervical esophagus. Webs were found in 5.5% of the cases, but only six patients had dysphagia attributable to the same, and none of the patients fulfilled the criteria for PVS.¹ In our case, the patient satisfied all the criteria of PVS.

Three to 15% of patients with PVS,³ mostly women between 15 and 50 years,¹ have been reported to develop esophageal or pharyngeal cancer. Paterson stated that post-cricoids

carcinoma was not an uncommon sequel of this condition.⁴ It is reported that iron and vitamin deficiency induced mucosal changes over a period become dysplastic and subsequently malignant.⁴

PVS can be treated effectively with iron supplementation and mechanical dilatation of the web(s).⁷ Mechanical dilation of webs is carried out by endoscopic dilatation in single/multiple sessions.⁷ Other means of disrupting an esophageal web are by neodymium-doped:yttrium aluminum garnet laser therapy or needle-knife electro incision. However, it is rarely used.

The patient should be maintained on a nutritious diet to establish the integrity of the oral epithelium since PVS is a potentially malignant disorder. The improvement was seen in the patient's swallowing after iron therapy provides sufficient evidence for an association between iron deficiency and post-cricoid dysphagia.⁸ The decline in PVS lately seems to parallel a betterment in nutritional status, including iron supplementation. Since post-cricoid webs in the PVS has been identified as a risk factor for the development of upper gastrointestinal tract malignancy, endoscopic surveillance, and regular follow-up is essential.^{9,10}

CONCLUSION

PVS remains a diagnostic challenge. Nevertheless, PVS should always be considered in a patient with dysphagia who also has associated features of iron deficiency anemia. Since it is a potentially malignant disorder, timely identification, treatment and monitoring of this syndrome is mandatory.

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