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Plummer-Vinson Syndrome. A Case Report

O. Hadadia[1*], M. Jarti[1], A. Ait Errami[1], S. Oubaha[2], Z. Samlani[1], K. Krati[1]

[1]Service de gastro-entérologie CHU Mohammed VI Marrakech
[2]Laboratoire de Physiologie, Faculté de médecine et de Pharmacie de Marrakech

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***Corresponding author:** Oualid Hadadia

Abstract **Case Report**

Plummer-Vinson syndrome (PVS) is characterized by a triad of symptoms comprising microcytic hypochromic anemia, esophageal webs, and dysphagia. PVS is commonly found in women of middle age especially in the fourth and fifth decade of life and is rarely reported in males. We report a case of a 53-year-old female patient who had a classic presentation of PVS. PVS is precancerous with high malignant potential; early diagnosis is of utmost importance for better prognosis and surveillance endoscopy is recommended. Iron repletion oftentimes improves the dysphagia; seldom esophageal dilatation is used to provide symptomatic relief.

Keywords: plummer vinson syndrome, iron deficiency anemia.

INTRODUCTION

Plummer-Vinson syndrome (PVS) is a rare condition characterized by the classic triad of dysphagia, iron-deficiency anemia, and esophageal web [1]. Plummer Vinson syndrome is more common in middle-aged women who appear to be at an increased risk of developing squamous cell carcinoma of the pharynx and proximal esophagus [2]. It is named after two American physicians Dr. Henry Stanley Plummer and Dr. Porter Paisley Vinson. PVS is also called Kelly-Paterson syndrome, named after two British otolaryngologists, Dr. Adam Brown-Kelly and Dr. Donald Ross Paterson [3]. Some differential diagnosis of PVS considered is dysphagia from achalasia, reflux esophagitis, esophageal carcinoma, esophageal spasm, systemic sclerosis, and Zenker's diverticulum [4]. Physical examination of patients with PVS is usually remarkable for pallor, glossitis, fatigue, and weakness [5]. Laboratory findings of PVS are consistent with the presence of iron deficiency anemia. Barium esophagogram is the best initial imaging study used for diagnosing PVS, which shows esophageal webs. Esophagogastroduodenoscopy (EGD) is also used to visualize esophageal webs and dilatation in a few cases [6]. The mainstay of treatment for PVS is aimed at correcting iron deficiency anemia.

CASE PRESENTATION

This is a 52-year-old Women with no particular pathological history, in particular no voluntary or involuntary intake of caustic products or notion of gastroesophageal reflux, no known food allergy or atopic site or notion of digestive or extra-digestive neoplasia or mediastinal pathology.

Admitted for aetiological assessment of a high dysphagia of organic appearance, painful, intermittent, evolving for 14 years, elective to solids respecting liquids of the type of attachment of the food bolus, without other associated digestive or extra-digestive manifestations, evolving in a context of apyrexia and maintenance of general condition, made up of asthenia and unstated weight loss.

The clinical examination noted a patient in good general condition with a WHO performance score of 0 and a BMI of 21.1 kg / m2. Blood pressure was 110/60 mmHg, heart rate 74 beats per minute and respiratory rate 18 cycles per minute.

The remainder of the physical examination was unremarkable apart from mucocutaneous pallor associated with signs of malnutrition such as melting of the adipose panniculus.

The biological assessment objectified a microcytic hypochromic anemia with a hemoglobin level at 10.5 g / dL, an average blood volume at 60.2 fL, the mean corpuscular hemoglobin concentration at 25 g / dL, white blood cells at 4900 uL and a level of platelets at 276,000 uL, low ferritinemia at 7 ng / mL

(N = 20-200), ionogram and renal function without abnormalities, as well as normal inflammatory workup.

The anti-transglutaminase antibodies (Ig G and Ig A) were negative. An eso-gastro-duodenal fibroscopy had revealed a diaphragm at the level of the mouth of the esophagus that could not be crossed with the endoscope, with cervical CT, a stenosing wall thickening, eccentric of the initial part of the esophagus, extended over approximately 3 cm without circumscribed tissue damage or visible peri-esophageal infiltration.

An eso-gastro-duodenal transit revealed a circular narrowing in regular diaphragm of the cervical esophagus located opposite C6-C7 compatible with Plummer-Vinson syndrome

Hypo-pharyngoscopy had demonstrated the presence of an impassable stenosis of the mouth esophagus with a central opening measuring 1 mm, without visible tumor. These arguments led to the diagnosis of Plummer-Vinson syndrome (Figure1).

As treatment, the patient received ferric iron in the amount of 100 mg. She then benefited from several sessions of dilations of the esophagus using SavaryGilliar candles and by means of a pneumatic balloon with progressively increasing diameters of 15 mm / 16 mm / 18 mm.

The endoscopic control visualized a dilaceration at the level of the dilated area with a good passage of the endoscope without jumping. Exploration of the remainder of the mucosa revealed a macroscopically normal esophageal mucosa, gastric and duodenal mucosa as well as the duodenal folds without abnormality. The course after four dilation sessions was favorable with resolution of the dysphagia. She is currently asymptomatic.

DISCUSSION

PVS was suspected in this patient with iron deficiency anemia and dysphagia; it was then confirmed after her barium swallow. The syndrome is a rare disorder characterized by a triad of iron-deficiency anemia, dysphagia, and esophageal webs [7]. The exact pathogenesis of PVS and the formation of the esophageal web is not well known. It has been postulated that iron deficiency induces iron-dependent enzyme dysfunction, leading to oxidative stress and DNA damages in the epithelia of the esophageal mucosa [2]. Repeated injury to epithelia due to iron deficiency leads to atrophy of mucosa and degradation of pharyngeal muscles, leading to the development of esophageal webs [8]. Patients with iron deficiency have low levels of myoglobin which may affect the muscles of the tongue and lead to glossitis. Genes involved in the pathogenesis of iron deficiency anemia associated with PVS include a mutation in the TMPRSS6 gene [9-10]. The TMPRSS6 gene encodes instructions for the protein hepcidin. Increased levels of hepcidin lead to decreased release of iron from ferritin and subsequently presents as iron deficiency anemia [11]. The role of mucosal inflammation and atrophy especially in the post-cricoid region has been suggested as a factor for the pathogenesis of PVS. The post-cricoid region experiences maximum trauma during swallowing of the solid bolus, leading to an increased risk of web formation [12, 13]. Histologically, webs in patients with PVS show fibrosis, epithelial atrophy, epithelial hyperplasia and hyperkeratosis, basal cell hyperplasia, and some features of chronic inflammation [14]. Esophageal webs and esophageal strictures are characteristic findings of PVS on the gross pathology. Microscopy can be notable for epithelial atrophy, chronic submucosal inflammation, and epithelial atypia or dysplasia in advanced cases. Medical therapy involves replacing the iron stores which can decrease the dysphagia symptoms effectively. Surgery is not the routinely recommended treatment option for patients with PVS [15-16]. In case of significant esophageal obstruction by multiple esophageal webs or persistent dysphagia despite medical treatment, rupture and mechanical dilation of the web using an endoscope can be performed such as with our patient [16].

CONCLUSIONS

It is important to further evaluate any complaint of dysphagia and to be cautious of any red flag warning signs. Especially in the setting of glossitis on the exam and severe iron deficiency anemia on labs, consider evaluating for esophageal webs as the etiology of dysphagia. Managing the esophageal webs of PVS and repleting iron stores at an early stage is crucial as the webs can progress to esophageal or pharyngeal squamous cell carcinoma.

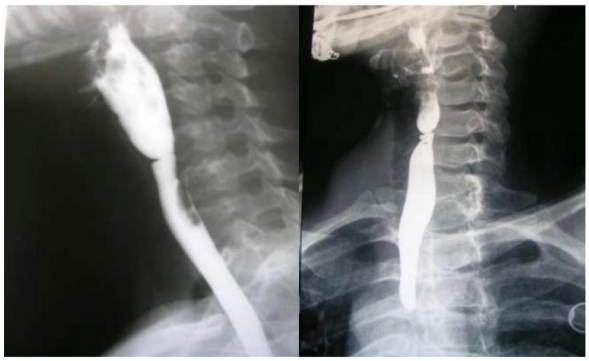

Fig-1: Appearance of diaphragm narrowing of the cervical esophagus on TOGD

Human Ethics

Consent was obtained by all participants in this study

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