Scholars Journal of Applied Medical Sciences
Abbreviated Key Title: Sch J App Med Sci
ISSN 2347-954X (Print) | ISSN 2320-6691 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Gastro-Enterology

Plummer-Vinson Syndrome: A Rare Cause of Dysphagia

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DOI: 10.36347/sjams.2021.v09i09.003 | **Received:** 25.07.2021 | **Accepted:** 30.08.2021 | **Published:** 04.09.2021

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Abstract **Case Report**

Plummer Vinson syndrome is a rare condition characterized by cervical dysphagia associated with iron deficiency anemia and a ring on the upper esophagus. Sometimes its unusual presentation can lead to misdiagnosis. We report the observation of a patient with Plummer-Vinson syndrome, revealed by upper dysphagia.

Keywords: Plummer-Vinson, iron deficiency anemia, dysphagia upper, esophageal diaphragm.

INTRODUCTION

Plummer-Vinson syndrome (PVS) is defined by a classic triad of iron deficiency anemia, upper dysphagia and the presence of an upper esophageal diaphragm. The exact etiology remains controversial but has been associated with nutritional deficiency, autoimmune disorders, hereditary factors, and a remarkably high female predominance. Dysphagia is the main symptom that can reveal it. However, some unusual presentations without dysphagia can misdiagnose. The risk of degeneration into squamous cell carcinoma in the pharynx and esophagus requires its recognition and early management. This article reports a presentation of Plummer-Vinson syndrome in a 50-year-old woman who consulted in the gastro-entero-hepatology department at the Arrazi hospital of the Mohammed VI University Hospital in Marrakech.

OBSERVATION

This is a 50-year-old patient 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 10 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 17.2 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 9.6 g / dL, an average blood volume at 60.2 fL, the mean corpuscular hemoglobin concentration at 25 g / dL, white blood cells at 8910 uL and a level of platelets at 285,000 uL, low ferritinemia at 2 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 (Figure 1).

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 (Fisure 2).

Hypo-pharyngoscopy had demonstrated the presence of an impassable stenosis of the mouth of the

esophagus with a central opening measuring 1 mm, without visible tumor. These arguments led to the diagnosis of Plummer-Vinson syndrome.

As treatment, the patient received ferric iron in the amount of 100 mg. He then benefited from several sessions of dilations of the esophagus using Savary-Gilliar 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.

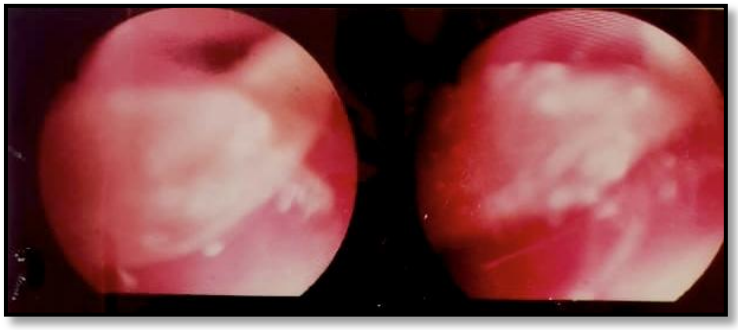

Fig-1: Endoscopic aspect of the membrane in the upper part of the esophagus

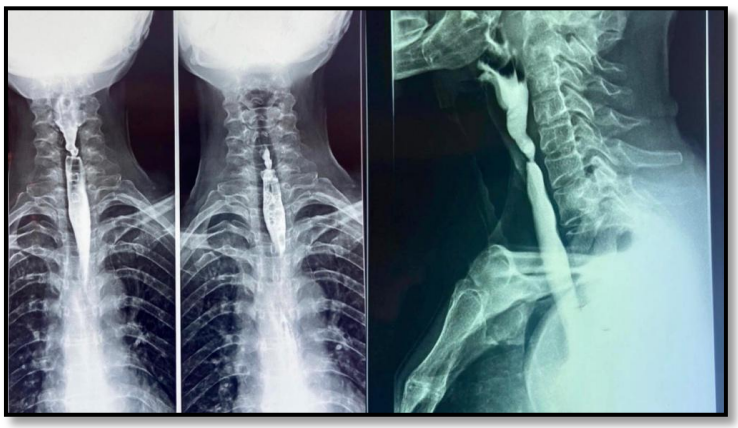

Fig-2: Appearance of diaphragm narrowing of the cervical esophagus on TOGD

DISCUSSION

The prevalence of Plummer-Vinson syndrome is not well established. This can be explained by the scarcity of world publications on this subject. SPV has been frequently reported in Northern Europe, particularly in rural Sweden [1]. Currently, it is increasingly described in sub-Saharan Africa where iron deficiency and malnutrition are common. The majority of cases occur in middle-aged women and are believed to be secondary to blood loss due to pregnancy and menstruation. Plummer-Vinson syndrome is characterized by the classic triad of dysphagia, iron deficiency anemia and a membrane in the proximal part of the esophagus [2].

Clinically, dysphagia is the main symptom. It is generally painless and intermittent, limited to solids. It is a rare condition that usually resolves with oral iron treatment, but it is not uncommon for endoscopic dilations to be required for its treatment [3]. Generally, the prognosis is good. A risk of squamous cell carcinoma of the esophagus has been described, which worsens the prognosis, therefore close monitoring should be carried out [4].

Several etiologies have been proposed [5]: iron deficiency, general nutritional deficiencies and gastric lesions. The syndrome has also been linked to autoimmunity and thyroiditis [6]. Links to genetically predetermined gastric atrophy have been suggested [7] but are not widely accepted. Wynder and Fryer [8] believed that the patients' diet was deficient in fresh fruits, vegetables and meats. Iron deficiency has been pathogenously linked to esophageal webs in that the esophageal lining is susceptible to iron deficiency, due to high cell turnover, thus causing rapid loss of iron-dependent enzymes leading to mucosal degeneration [9]. It has been suggested that such continuous tissue damage and scarring causes permanent changes in the esophageal lining [10].

The mechanism of dysphagia and membrane formation is not well understood. Thus, even if iron deficiency is not necessary for the formation of the membrane, it would always precede dysphagia. However, the membrane is not thought to be pathognomonic in SPV [11]. It would not, in fact, be necessary for the onset of swallowing disorders but rather play a worsening role of a mechanical nature. But iron deficiency due to pernicious anemia seems to be the most logical cause. The usual haematologic expression of VPS is microcytic hypochromic anemia with lowered ferritinemia associated with stigmata of malabsorption.

Usually, the seat is more often posterior; the ring is single but can be multiple. The histologic appearance is generally mucosal atrophy with a polymorphic inflammatory submucosal infiltrate associated with atrophy of the muscular mucosa [12]. Treatment is based on iron supplementation. In most cases, this would lead to regression of dysphagia even before the biological normalization of anemia, despite the persistence of the esophageal ring. However, according to Jones [13] an obstructive ring is often the cause of resistance to treatment requiring endoscopic treatment with candle dilation sessions. Short-term developments are quickly favorable. A recurrence is possible especially in the event of new iron deficiency. In our patient, after receiving a blood transfusion and an infusion of injectable iron, endoscopic dilation was performed under balloon sedation with good progress without recurrence to date. In the long term, the course is marked by an increased risk of post-cricoid cancer, which makes SPV a precancerous condition. Several series have reported an incidence of 3 to 15% of esophageal and post-cricoid carcinomas [12]. Therefore, annual endoscopic monitoring is recommended.

CONCLUSION

Plummer Vinson syndrome is a rare cause of mechanical dysphagia. Intermittent dysphagia, young age and male sex can misdiagnose. The increased risk of degeneration should motivate close endoscopic monitoring. The increased risk of degeneration should motivate close endoscopic monitoring with biopsies.

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