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Original Research Article

Celiac Disease Associated with Plummer-Vinson Syndrome

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Abstract

Introduction: Plummer-Vinson syndrome (PVS), also known as sideropenic dysphagia, is a very rare entity that combines a classic triad: iron deficiency anemia, dysphagia and upper esophageal diaphragm. Its association with celiac disease has been rarely reported. The goal of this work is to determine the clinical characteristics of patients with PVS, in celiac disease, thus the evolutionary profile. ***Materials and Methods:*** This is a descriptive retrospective study covering a period of 26 years, from January 1993 to January 2019, collecting all patients followed in the service of Medecine C at the University Hospital Ibn Sina of Rabat-Morocco for PVS who have systematically benefited from jealousy biopsies in search of celiac disease. ***Results:*** Out of a total of 149 patients followed in the PVS unit, the prevalence of celiac disease was 6.1% (10 cases). 8 cases diagnosed as part of the etiological assessment of PVS, the diagnosis of celiac disease was concomitant with PVS in 2 cases. These were 8 women and 2 men with a sex ratio of 0.25. The average age was 28 years [19- 56 years]. All patients had organic dysphagia, five cases (62.5%) had clinical anemic syndrome associated with malabsorption diarrhea. Upper GI fibroscopy showed a ring at the killian's mouth in all cases. After oesophageal dilation by candles of different diameters, fibroscopy showed a rarefaction of duodenal folds in 9 cases (90%) and a duodenum of normal appearance in one case (10%). The anatomopathological study of duodenal biopsies showed intraepithelial lymphocytosis (IEL) > 30% in all cases, moderate atrophy of the villus in 6 cases and severe atrophy in 4 patients. All our patients have received martial treatment in combination with a gluten-free diet (GFD). The progression was favourable in 8 patients after a single dilation session and a well-followed GFD; 2 patients with poor GFD compliance also had a recurrence of dysphagia. ***Conclusion:*** PVS on celiac disease remains rare, found only in 3.5% in our series; good compliance with the GFD has improved signs of malabsorption and the disappearance of dysphagia in 80%.

Key words: Celiac Disease, Vinson Syndrome, sideropenic dysphagia.

INTRODUCTION

Plummer-Vinson syndrome (PVS), also known as Kelly-Paterson syndrome or sideropenic dysphagia, is characterized by the association of a triad: dysphagia, sideropenic anemia and diaphragm or esophageal "Web". The association of this syndrome with celiac disease or food intolerance to gluten is uncommon. The association of these two diseases was first reported by Dickey and McConnell in 1999 [1].

The goal of this work is to determine the clinical characteristics of patients with PVS with celiac disease, as well as the evolutionary profile under a gluten-free diet (GFD).

MATERIALS AND METHODS

This is a descriptive retrospective study covering a period of 26 years, from January 1993 to January 2019, collecting all patients followed for PVS in the Department of Medecine C at the University Hospital Ibn Sina, Mohamed V University, Rabat-Morocco; and in whom the diagnosis of associated celiac disease has been made.

All our patients have benefited from dilatation and jejunal biopsies.

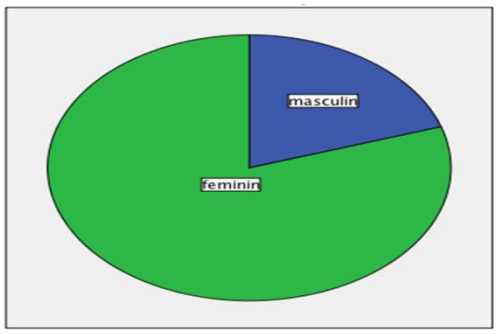
The statistical analysis of the data was done using the SPSS 21 software.

RESULTS

Out of a total of 149 patients followed in the PVS unit, the prevalence of celiac disease was 6.1% (10 cases). Eight cases diagnosed as part of the etiological assessment of PVS. The diagnosis of celiac disease was concomitant with in 2 cases.

The average age was 28 years [19- 56 years].

These were 8 women and 2 men with a sex ratio of 0.25 (Graph: 1)



Distribution by Sex

Five cases (50%) had a history of microcytic hypochrome anemia when martial treatment was started, one case (10%) was followed for HTP on HAI type I, one case (10%) was followed for HTP on post-viral cirrhosis C (CPVC), and one case (10%) had ATCD of deep vein thrombosis of the lower limbs.

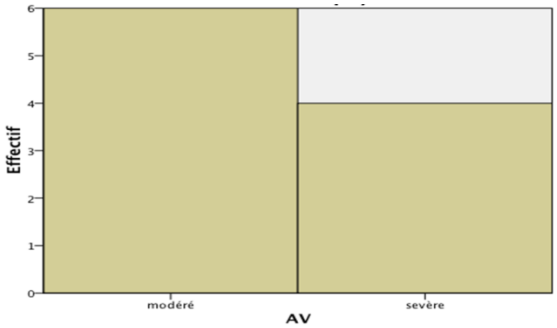
All patients had organic dysphagia, five cases (62.5%) had clinical anemic syndrome associated with malabsorption diarrhea, and one case had odynophagia.

The diagnostic time for VPD in relation to the occurrence of dysphagia was on average 4.3 ±5.3 years.

All patients received at least 1 session of esophageal dilation by progressive diameter Guillard savary candles.

Upper GI fibroscopy before dilation showed a ring at the killian mouth in all patients, and after endoscopic dilation with rupture of the ring, upper GI exploration revealed esophageal varices in 2 cases (20%), rarefaction of duodenal folds in 9 cases (90%) and a duodenum of normal appearance in one case (10%).

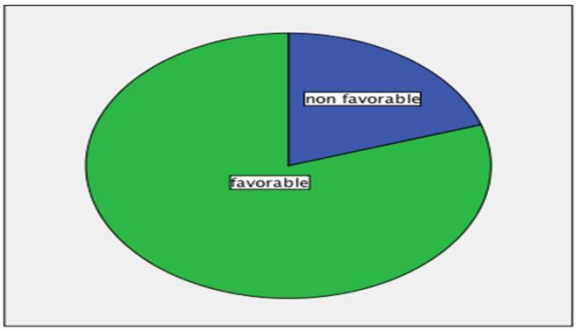
The anatomopathological study of duodenal biopsies showed intra2pithelial lymphocytosis (IEL) > 30% in all cases, moderate atrophy of the villus in 6 cases and severe atrophy in 4 patients (graph 2).



Anatomopathological data
Villous atrophy

All patients received at least 1 session of esophageal dilation by progressive diameter Guillard savary candles, martial treatment in combination with a gluten-free diet (GFD) explained by a cell of dieticians.

In our series, the progression was favourable in 8 patients after a single dilatation session and a well followed GFD; 2 patients with poor GFD compliance also had a recurrence of dysphagia (Figure 2). The GFD has led to clinical and histological improvement. Indeed, the control intraepithelial lymphocytosis was less than 30% with total restitution of villosities in all 8 cases.



Evolutionary data
Disappearance or recurrence of dysphagia

DISCUSSION

PVS, also known as Paterson-Brown-Kelly syndrome or Kelly-Paterson syndrome or sideropenic dysphagia, is characterized by the association of an esophageal diaphragm responsible for sideropenic anemia dysphagia. This syndrome was first described by Henry Plummer in 1912 [2] and then by Vinson in 1919 [3]. It is a rare entity [4], affecting mainly women between 40 and 70 years of age [5-6], but this syndrome has also been described in men, children and adolescents[7-8]. This syndrome is most commonly described in northern European countries, particularly in rural Sweden[9]. In Africa, although malnutrition and iron deficiency are more prevalent, this condition was described until recently when it is increasingly described [4, 10-12].

The etiopathogenesis of this syndrome remains unknown [4,13]. The association with sideropenic anemia, which is one element of the triad of definitions of this syndrome, suggests an important etiological role of this anemia in the genesis of the esophageal diaphragm. Also, some studies have shown that the mere correction of this anemia in some patients may lead to an improvement in dysphagia [9]. Other etiopathogenic factors may be involved in the genesis of PVS, such as genetic, immunological and environmental factors, which explain the association with certain autoimmune diseases [14, 13,15].

Clinically, PVS is characterized by the presence of dysphagia, which is generally painless and intermittent or slowly progressive, selective for solids and sometimes associated with weight loss. Clinical anemic syndrome (pallor, generalized weakness, fatigue, tachycardia, dizzying sensations) may sometimes dominate the clinical picture. Other signs described are glossitis, angular cheilitis and koilonychia[4]. Dysphagia, often post-cricoid, during PVS is related to the existence of a semilunar membrane of the cervical esophagus (membranous fold below the pharyngeal-esophageal junction) which is often highlighted on esophageal digestive opacifications as a short, suspended stenosis, in diaphragm or ring, facing the cervical vertebrae (C5-C6) [16]. This diaphragm can also be detected during upper digestive endoscopy such as annular, fibrous stenosis, with a central or sometimes eccentric lumen. It usually sits in the upper third of the esophagus and may be misunderstood or accidentally broken during rapid introduction of the endoscope [2, 4,5].

The management of a patient with PVS begins with the search for a possible cause of iron deficiency anemia. This investigation begins with the exclusion of a lack of intake, bleeding, neoplasia, celiac disease or gluten intolerance [4]. Indeed, although the association of PVS and celiac disease is uncommon, the latter, when present, is a common cause of martial deficiency by intestinal malabsorption, responsible for iron deficiency or sideropenic anemia. This association of PVS with celiac disease was first reported by Dickey and McConnell in 1999 [1]. The diagnosis of celiac disease is based on clinical, biological and histological arguments. The clinical picture is usually variable, from so-called "dry" forms to polydeficient diarrhoeal forms. In biology, while iron-deficiency anemia can classically reveal celiac disease, its poly-deficiency origin (iron, folates) reflects its often dimorphic nature, with an average globular volume remaining within normal limits. Serology usually shows the presence of anti-endomysium or anti-transglutaminase IgA antibodies. Duodenal biopsies confirm the diagnosis by showing villous atrophy associated with lympho-plasmocytic infiltration of the chorion and an increase in intraepithelial lymphocytes on histopathological examination above 30%. All these lesions can regress under a gluten-free diet [17].

Celiac disease, especially when the gluten-free diet is not or poorly followed, increases the risk of intestinal lymphoma, but also of other cancers, in particular squamous cell carcinomas of the pharynx and oesophagus [18]. In addition, there is a risk of esophageal cancer increased in the long term by the pre-existence of an PVS, which is thus a pre-neoplastic condition [19,20]. This justifies regular follow-up in this group of patients, such as our patients, given the risk of developing carcinoma of the oesophagus or pharynx.

PVS can complicate an untreated or poorly treated celiac disease, as it can precede it, sometimes by several years [1]. In our series, celiac disease was already known in 2 cases.

CONCLUSION

The association of PVS with celiac disease is uncommon, and remains rare, found only in 3.5% in our series. However, in any patient with VPS, celiac disease should be discussed and investigated, especially if the cause of the anemia is not obvious. The treatment of these patients is based on parallel management including correction of deficiencies, a gluten-free diet and often esophageal dilation, with a favourable course in most patients; good compliance with the GFD has improved the signs of malabsorption and the disappearance of dysphagia in 80%. In patients with VPS associated with celiac disease, regular endoscopic monitoring should be maintained in view of the risk of developing carcinoma of the esophagus and pharynx.

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