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Case Report

Plummer-Vinson Syndrome: A Case Report

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Abstract: Plummer-Vinson syndrome (PVS) is a rare disease characterized by cervical dysphagia associated with iron-deficiency anemia and an upper esophagus web. Sometimes, its uncommun presentation may lead to misdiagnosis. An upper gastrointestinal endoscopy may incidentally reveal annular stricture. We report a case of a young patient with Plummer-Vinson syndrome during an upper digestive symptomatology manifested by isolated dysphagia. The diagnosis of Plummer-Vinson syndrome was confirmed by the etiological investigation and its treatment consisted of iron supplementation and endoscopic dilation, resulting in a faster recovery.

Keywords: Iron-deficiency anemia, upper dysphagia, esophageal ring/web, Plummer-Vinson syndrome.

INTRODUCTION

Plummer-Vinson syndrome, also known as Patterson-Kelly syndrome, is characterized by a classic triad comprising dysphagia, sideropenic anemia and the presence of esophageal membranes. Dysphagia remains the main symptom of the syndrome. However, some uncommun presentations without dysphagia [1] may lead to misdiagnosis. However, the risk of undergoing degeneration into squamous cell carcinoma in the pharynx and esophagus must be recognized and requires early management. We report an observation of Plummer-Vinson syndrome, discovered in the course of an upper digestive symptomatology during an upper digestive symptomatology manifested by isolated dysphagia in a young patient at Mohamed VI University Hospital (CHU) of Marrakech..

OBSERVATION

The case involved a 20-year-old patient who had no particular pathological antecedents, namely voluntary or involuntary intake of caustic substances, Gastro-oesophagien Reflux (GERD), common food allergy. She also had no atopic predisposition nor digestive or extradigestive neoplasia, nor ENT pathology nor any common mediastinal pathology. He got an appointment for an organic high dysphagia which had been developing for 2 years and causing ongoing pain. The patient experienced problems swallowing solid foods but had no trouble with liquids and showed no other associated digestive or extradigestive manifestations. Overall, he was apyrexial and in good physical condition. The physical examination revealed that the patient was in good condition with a WHO performance status of 0 and a BMI of 20.2 kg / m2. Blood pressure was 100/70 mmHg, heart rate was 75 beats per minute and respiratory rate was 17

breaths per minute. The patient had a clear cutaneous-mucous pallor, without other abnormalities of skin appendages. The rest of the physical exam was normal. Esophagogastroduodenal fibroscopy was performed and revealed the presence of a diaphragm in the upper esophageal sphincter that proved impassable for the endoscope (Figure 1).

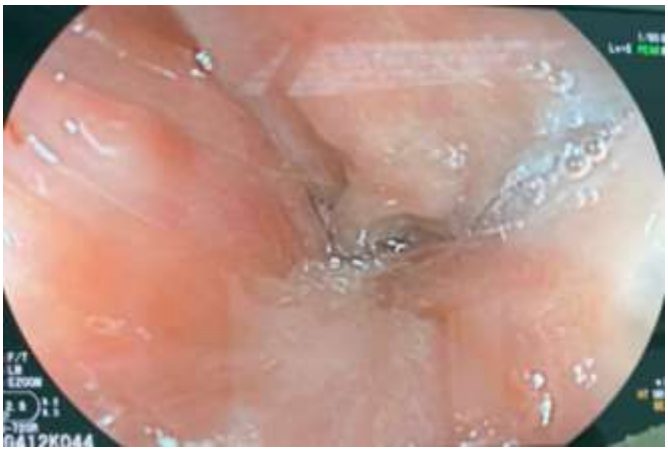

Fig-1: Endoscopic aspect of the ring at the level of the esophgeal entrance

The CT scan detected stenosing esophageal wall thickening, eccentric from the esophageal entrance and extending approximately 3 cm without a circumscribed mass lesion or peri-esophageal infiltration.

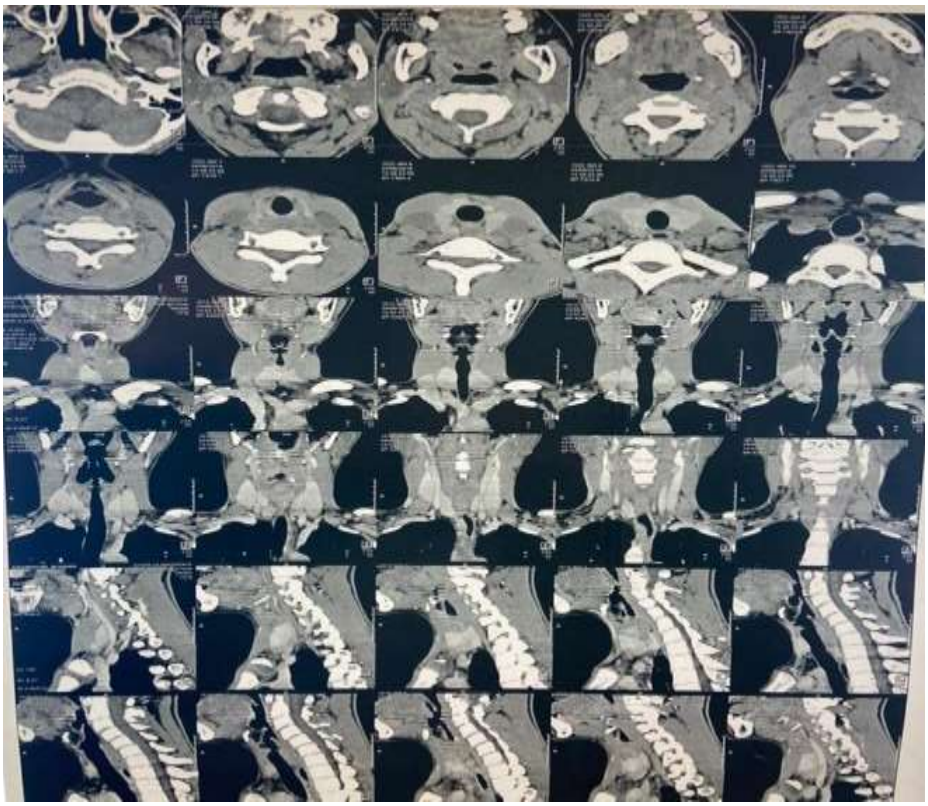

Fig-2: CT appearance of esophageal stenosis: Stenosing wall thickening, eccentric of the esophagus entrance

Esogastroduodenal transit showed esophageal stenosis compared to C5-C6 with upstream stasis, most likely related to a stenosing diaphragm (Figure 2).

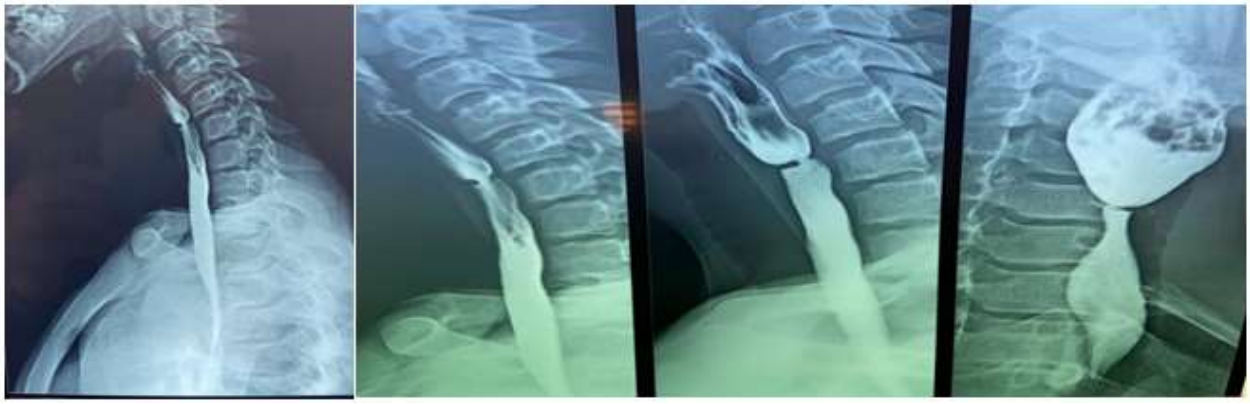

Fig-2: Imaging of oeso-gastro-duodenal transit (OGDT) stenosis

At the biological level, the blood count revealed hypochromic microcytic anemia with a hemoglobin level of 6.8 g / dl, a mean corpuscular volume of 53.4 um3 (N = 78 -98), a mean corpuscular hemoglobin concentration of 26.2 g / 100ml (N = 31-36.5), a mean corpuscular hemoglobin of 14pg (N = 26-34). White blood cells counts were 3510 / mm3 and platelets 367,000 / mm3. The iron reserves were totally depleted with a ferritin <1 ng / ml (N = 20-200). In total, it was a painful dysphagia with a stenosing diaphragm at the level of the esophagus entrance, associated with iron-deficiency anemia, suggesting the diagnosis of Plummer-Vinson syndrome. The patient received a red blood cell transfusion and intravenous iron infusion. After achieving an acceptable hemoglobin level> 8g / dL, the patient underwent esophageal dilation under sedation by means of a balloon with gradually increasing diameters of 15mm / 16mm / 18mm. The endoscopic control visualized a laceration in the mucosa of the upper part of the esophagus and a smooth passage of the endoscope.

As part of the etiological assessment of his iron-deficiency anemia, we performed:

- An esophagogastroduodenoscopy (EGD) with duodenal biopsy which revealed an aspect of erythematous pangastritis with preserved duodenal folds. The anatomicopathological exam indicated a subacute and chronic non-specific duodenitis with moderate chronic pangastritis of moderate to severe activity, moderately atrophic without metaplasia or dysplasia with the presence of Helicobacter pylori.
- A total colonoscopy with catheterization of the last ileal loop which revealed a granitized ileal mucosa and whose anatomopathological examination indicated a subacute and chronic non-specific ileitis with subacute and chronic non-specific inflammatory change of the colorectal mucosa without sign of malignancy.
- Celiac disease was excluded due to IgA anti-transglutaminase antibodies which were found negative with an IgA dosage of 2.9 g / L (N = 1.1 - 3.6).

The etiology of iron deficiency was likely attributable to atrophic Helicobcater pylori gastritis which was eradicated by concomitant quadruple therapy. The clinical and biological course was satisfactory, the patient no longer suffers from dysphagia and to date, after a 7-month follow-up, iron deficiency has not recurred.

DISCUSSION

The prevalence of Plummer-Vinson syndrome (PVS) is not well established. This is explained by the scarcity of global publications on the subject. PVS has been frequently reported in Northern Europe, particularly in rural areas in Sweden [2]. Currently, it is increasingly described in sub-Saharan Africa where iron deficiency and malnutrition are common. PVS is seen primarily in white women aged 30 to 70 [2]. Our patient's earlier age of onset differs from cases reported in the West. In Africa, published case series report an average age of 39 with extremes ranging from 20 to 65 years [2]. Clinically, dysphagia is the main symptom. It is generally 75% painless and intermittent and limited to solids [2]. In our patient, dysphagia was painful and had been going on for 2 years causing difficulty swallowing solids but not liquids. In PVS, the mechanism of dysphagia and membrane formation are still not well understood. So, even if iron deficiency is not necessary for ring formation, it would always precede dysphagia. However, the membrane would not be pathognomonic of PVS [3]. It is not in fact necessary for the onset of swallowing disorders, but rather plays a role of mechanical aggravation. But, iron deficiency due to pernicious anemia seems to be the most logical cause. The common haematological expression of PVS is microcytic hypochromic anemia with

hypoferritinemia associated with signs of malabsorption. ROBERTO *et al*. [4] studied the motor function of the esophagus in patients with PVS and concluded that there was a movement disorder in the form of a decrease in the amplitude of contractions and disorders of their propagation due to the deficiency in serum iron. Anatomically, there is a membranous ring in the 1/3 upper esophagus below the cricopharyngeal muscle [2].

Esophageal rings are usually located in the distal esophagus and exist as a single lesion but can also be multiple. The histologic appearance is generally mucosal atrophy with a polymorphic inflammatory submucosal infiltrate associated with mucosal muscle atrophy [5]. 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 [6], an obstructive ring is often the cause of resistance to treatment necessitating endoscopic treatment consisting of bougie dilation sessions although the short-term course proves quickly favorable. As we could see it in our patient with a totally obstructive ring, after a single session of esophageal dilation associated with the correction of iron deficiency and anemia, there was no recurrence. The course was marked by an increased risk of post-cricoid cancer, which makes PVS a precancerous condition. Several series have reported an incidence of between 3 and 15% of esophageal and post-cricoid carcinomas [5]. Therefore, annual endoscopic monitoring is recommended.

CONCLUSION

Plummer-Vinson syndrome is a rare cause of mechanical dysphagia. The wide distribution of upper digestive endoscopy for any iron-deficiency anemia would allow a new approach to assessing its prevalence. The increased risk of a neoplastic graft should motivate close endoscopic monitoring with biopsies

REFERENCES

1. Elwood, P.C., Jacobs, A., Pitmann, R.G. (1964). Epidemiology of the Patterson KeIly syndrome. *Lancet, 3;2*(7362); 716-20.
2. Beggar, H., Benzzoubeir, N., Errabih, I., Ouazzani, L., Ouazzani, H. (2017). Syndrome de Plummer Vinson : A propos de 16 cas. Revue Marocaine des Maladies de l'Appareil Digestif; 25.
3. Louis Michel Wong Kee Song. (2008). Plummer Vinson syndrom treatment and medication gastro Mayo clinic.
4. Yasawy, M. I. (2004). Treatment of Plummer-Vinson syndrome with Savary-Gilliard dilatation. *Saudi medical journal*, *25*(4), 524-526.
5. Ben, Gamra, O., Mbarek. (2007). Syndrome de plummer Vinson. *la tunisie médicale, 85*(5); 402-404
6. Patterson, D.R. (1919). *Journal of laryngology,* 34; 289.