Plummer-Vinson syndrome

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RÉSUMÉ

Prérequis : Le syndrome de Plummer-Vinson est une affection rare, qui atteint essentiellement la femme de race blanche. Il se caractérise par une dysphagie, une carence martiale et la présence d'un anneau membraneux du tiers supérieur de l'œsophage.

But : Le but de cette étude était de préciser les caractéristiques épidémiologiques, cliniques, para cliniques, thérapeutiques et évolutives du syndrome de Plummer-Vinson.

Méthodes : Nous rapportons 10 cas de syndrome de Plummer-Vinson, colligés sur une période de 6 ans (2002-2008). Les paramètres étudiés étaient l'age, le sexe, les signes cliniques et biologiques, notamment la numération de la formule sanguine, l'albuminémie, les bilans hépatique, rénal et lipidique. Tous les patients étaient explorés par une endoscopie digestive haute.

Résultats : La dysphagie était le principal symptôme, retrouvé dans tous les cas. L'anémie sidéropénique était observée chez 50% des patients. La prise en charge thérapeutique était basée sur la dilatation endoscopique, qui a été réalisée chez tous les patients, dont 3 ont nécessité plusieurs séances, et sur le traitement martial qui n'était prescrit qu'en cas d'anémie. L'évolution initiale était favorable dans tous les cas avec une disparition de la dysphagie. Deux cas de syndrome de Plummer-Vinson était découverts au stade de dégénérescence.

Conclusion : La dysphagie constitue le principal symptôme pouvant révéler un syndrome de Plummer-Vinson. La dilatation endoscopique est une procédure de choix dans le traitement des diaphragmes oesophagiens au cours de ce syndrome. Le risque de dégénérescence impose une surveillance endoscopique régulière.

Mots-clés

Syndrome de Plummer–Vinson – Dysphagie - endoscopie haute

SUMMARY

Background : Plummer – Vinson syndrome is one of the names given to the constellation of dysphagia, iron- deficiency anemia, and esophageal webbing. It is a rare affection wich affects mainly white women.

Aim: The purpose of this study is to precise epidemiological, clinical, paraclinical and therapeutic features of the affection.

Methods: We report a retrospective study enrolled over 6 years (2002 - 2008). Were studied the age, the sex, the main symptoms and the biological parameters (full blood cell, albumin, hepatic, renal and lipidic balance sheet). Patients were explored by an upper endoscopy.

Results: Ten patients with the diagnosis of Plummer-Vinson syndrome were collected. There were 9 women, the average age was 57 years old. Dysphagia was the main symptom, observed in 100% of the cases. Fifty per cent of our patients had iron-deficiency anemia. Iron supplementation was indicated each time there is an iron- deficiency anemia. All the patients were treated with endoscopic dilatation and three of them needed multiple session of endoscopic dilatation. Eighty per cent had a favorable evolution. The malignancy was observed in 2 cases.

Conclusion: Dysphagia is the main symptom of the Plummer–Vinson syndrome, which must indicate an upper endoscopy. This syndrome is known to be associated with an increased risk of squamous cell carcinoma of the upper airway tract, so the patients should be followed closely. Endoscopic dilatation is the procedure of choice in the treatment of cervical web of the esophagus.

Key-words

Plummer –Vinson syndrome – Dysphagia - Upper endoscopy

متلازمة بلامر - فانسون : تجربة قسم أمراض المعدة و الجهاز الهضمي

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الكلمات الأساسية : متلازمة بلامر فانسون - عسر البلع - تنظير علوي

Plummer-Vinson syndrome, also known as Paterson Kelly syndrome and sideropenic dysphagia, refers to the constellation of dysphgia, iron-deficiency anemia and esophageal webs. It has been known since the beginning of the 20th century. Plummer [1] established the syndrome for the first time in 1912 and has published 21 cases with diffuse dilatation of the esophagus and spasm of the upper esophagus without anatomic stenosis. Then Vinson [2] Plummer's pupil, published another case with angulation of the esophagus. Paterson and Kelly [3-7] described for the first time the clinical signs of the syndrome: anemia, dysphagia, glossitis, cheilitis, iron deficiency and koilonychias. The treatment of Plummer- Vinson syndrome is iron supplementation and endoscopic dilatation. The syndrome is associated with an increased risk of upper aero-digestive tract carcinoma. Thus, the aim of the present study was to precise the epidemiological, clinical, paraclinical and therapeutic features of this affection.

PATIENTS AND METHODS

Patients

It is a retrospective study in which we reviewed the hospital medical records of 10 patients in whom the Plummer-Vinson syndrome was diagnosed at our center, from 2002 to 2008.

Methods

The diagnosis was established according to the endoscopic features. Several features were precised such as age, sex, past medical history, clinical presentation, endoscopic features and therapeutic management.

The data was summarized by descriptive statistics and analysed with SPSS version 10. Data are shown as mean ± standard deviation (SD). Differences in proportions were analysed by chi-square test; differences in mean quantitative value were analysed by student's t-test. P value less than 0.05 was accepted as statistically significant.

RESULTS

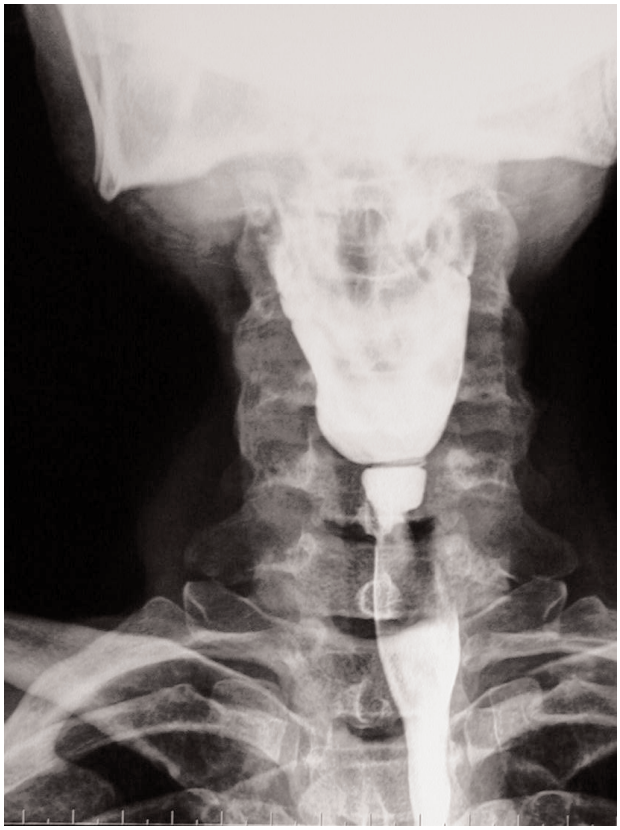
From the total sample, nine were women and only one was a man. The average age was 57 years old (limits: 30-75 years). A medical history of iron-deficiency anemia was found in five patients, among them one patient was diagnosed with celiac disease. One patient was diagnosed with iron-deficiency anemia while the Plummer-Vinson syndrome was established. Six patients among the nine women was menopaused while the diagnosis was established.

A slowly progressive dysphagia was the main symptom, observed in 100% of the cases. Sixty per cent of our patients presented with symptoms resulting from anemia (weakness, cutaneomucusal pallor, and tachycardia). Only one patient had angular cheilitis and koilonychias and one had a glossitis.

Hematological tests showed a low level of hemoglobin in 5 patients, which was ranging between 6.4 and 9 g/dl (average 10.6g/dl). Biological findings of iron deficiency (ferritinemia, plasmatic iron) were found in three patients. In one case repeated episodes of menorrhagia and in another case celiac disease resulted in depletion of the iron stores of the body.

The patient's esophagogram revealed the presence of cervical esophageal webs in 6 patients and a regular stenosis in 2 patients (Fig 1).

Figure 1 : Esophageal web in the barium swallow



Endoscopic examination was performed in 100% of our patients. The endoscope did not pass through at the level of the web in two times (Fig 2, 3).

Figure 2 : Endoscopic oesophageal web presentation

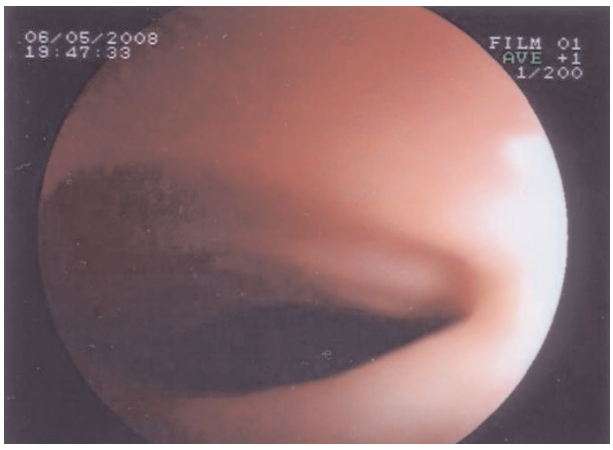
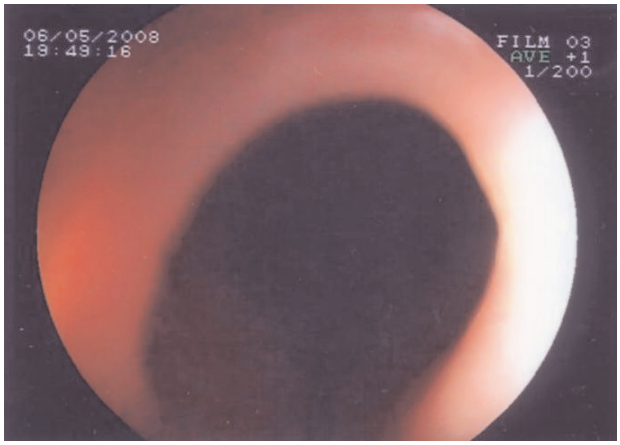
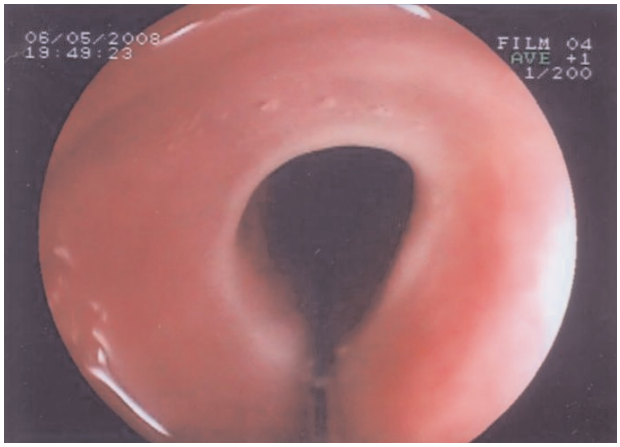


Figure 3 : Endoscopic oesophageal web presentation



The patients who presented with iron deficiency anaemia were treated with oral ferrous agent at the dose of 150 mg daily. Successful rupture of the webs was accomplished by the use of dilatation in 100% of our patients. Dilation was done by an endoscopic placement of guidewire dilators with a mean diameter of 11.7mm (limits: 7 to 15mm) (Fig 4). The illustration 5, showed the hemorrhage due to the web rapture by dilation. Three patients necessitated multiple session of endoscopic dilatation, two of them was found to have respectively an epidermoide carcinoma of the oesophagus and a carcinoma in situ of the oesophagus. No complication was recorded. The remaining 8 patients had no recurrence of symptoms in the follow up.

Figure 4 : Savary-Gilliard dilation in the management of cervical web of esophagus



DISCUSSION

In our study, the diagnosis of Plummer-Vinson syndrome was done in ten patients, 9 of them were women, with an average age of 57 years old. Dysphagia was the main symptom, observed in 100% of the cases. Fifty per cent of our patients had iron-deficiency anemia. Iron supplementation was indicated each time there is an iron- deficiency anemia. All the patients were treated with endoscopic dilatation and three of them needed multiple session of endoscopic dilatation. Eighty per cent had a favorable evolution.

Exact data about the incidence and prevalence of the Plummer-Vinson syndrome is not available [6]. In the past, this syndrome was more common among the Scandinavian population, but nowadays, it is extremely rare [5, 6].

The incidence has recently decrease because of nutritional improvement, advanced health care, a decrease incidence of pregnancy, improvement care program of pregnancy and better treatment of iron deficiency [5, 6]. In a series of 1000 patients who underwent a cineradiographic examination of the hypopharynx and the cervical esophagus, webs were found in 5.5% of the cases but only six patients had dysphagia due to the web and none of them fulfilled the criteria for Plummer-Vinson syndrome [8]. In Africa, where both iron deficiency and malnutrition are common, the syndrome is very rare [9]. Plummer-Vinson syndrome affects mainly white women (90%), in the fourth to seventh decade of life, and this group account for 75% in number [5, 7, 10], but some cases in children and adolescents have been reported [11, 12, 13]. In our study, 9 patients were women, and their mean age was 57 years old. The dysphagia is usually painless and intermittent or progressive over years, limited to solid and sometimes associated with weight loss. A slowly progressive dysphagia was noted in all our patients. Symptoms resulting from anemia such as weakness, pallor, fatigue and tachycardia may dominate the clinical picture [7, 14]. Additionally it is also characterized by glossitis, angular cheilitis and koilonychias [5]. Enlargement of the spleen and thyroid may also be observed [15, 16]. Sixty per cent of our patients presented symptoms resulting from anemia. The diagnosis is based on the evidence of iron-deficiency anemia and one or more esophageal webs in a patient with postcricoid dysphagia. Barium swallow X-ray can detect esophageal webs, but the best way for demonstration is the videofluoroscopy [14, 15, 17]. Webs are also detectable by upper gastrointestinal endoscopy. All our patients underwent an upper endoscopic examination. They appear smooth, thin and gray with eccentric or central lumen. The webs may be found in approximately 5-15% of patients with dysphagia [9]. It typically occur in the proximal part of the esophagus and may be missed and accidentally ruptured unless the endoscopes in introduced under direct visualization [15, 16]. Esophageal webs were found in 60% of the patients in our study, with a regular stenosis in 2 others patients. Laboratory examination typically reveal iron deficiency anemia with decreased values of hemoglobin, hematocrit, mean corpuscular volume, serum iron and ferritin and increase total iron binding capacity. In our study, only three patients had a biological finding of iron deficiency. Since dysphagia is a main clinical feature of Plummer-Vinson syndrome, the differential diagnosis includes all others causes of dysphagia especially malignant tumors, benign strictures or esophageal rings [5, 14].

The pathogenesis of this syndrome remains unclear, but

possible etiopathogenic mechanisms include iron deficiency, genetic predisposition or autoimmune disorder. Plummer-Vinson may be accompanied by pernicious anemia, thyroiditis, rheumatoid arthritis or celiac disease [5, 10, 15, 18]. In one of our patients, celiac disease was associated to the Plummer-Vinson syndrome.

For the management of the Plummer-Vinson syndrome, it is necessary to clarify the cause of the iron deficiency to exclude active hemorrhage, malignancy or celiac disease [14]. The treatment is based on iron supplementation, and iron therapy may be necessary even though hematologic parameters are normal in the presence of a web formation.

However, in case of significant obstruction of the esophagus lumen by esophageal web and persistence of dysphagia despite iron supplementation, rupture and dilatation of the web should be performed. After endoscopic placement of a guidewire, dilators with diameter up to 17 mm can be used [19]. Usually only one dilation is enough to relieve dysphagia, but occasionally multiple sessions are required. All our patients were treated by an endoscopic guidewire dilatation with no complications. Only three patients needed a multiple session of dilation. In addition, successful balloon dilation has been described [5].

This syndrome is associated with an increase incidence of postcricoid carcinoma (3-15%), so it is considered as a pre-cancerous lesion [20] and in rare cases of the stomach [7, 14]. A surveillance upper gastrointestinal endoscopy is recommended every year, even though the effectiveness of this recommendation is not definitively confirmed [15]. Prognosis of the Plummer-Vinson syndrome is excellent, unless it is associated with squamous cell carcinoma of the hypopharynx or upper esophagus which prognosis worsens dramatically.

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