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Research Article

Efficiency and Safety of Endoscopic Dilation of the Plummer-Vinson Ring

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Abstract

Introduction: The Plummer Vincent syndrome (PVS) or Kelly Paterson is a rare entity defined by the association of upper dysphagia, iron deficiency anemia and a fibrous ring of the cervical esophagus. Although correction of sideropenia may improve these symptoms, endoscopic dilatation of the esophageal diaphragm is often necessary. The main objective of our work is to analyze the results of endoscopic treatment and then to study the epidemiological, clinical and endoscopic characteristics of PVS.

Methods: We included in this study 77 cases of PVS collected in our unit between September 2005 and April 2018. All patients underwent martial treatment and endoscopic dilation of the esophageal ring with Gillard Savary Bougies or hydrostatic balloon under or without fluoroscopy. Other session (s) of dilatation was performed in case of recurrence of dysphagia and/or oesophageal stenosis under sedation with propofol.

Results: 11,373 upper gastrointestinal endoscopies were performed, including 184 in the context of PVS, over a period of 13 years (1,6%).

A total of 77 patients with PVS were included, including 66 women (85.7%) and 11 men (14.3%). The average age was 39 (range: 16 - 78 years). All patients presented dysphagia and anemic syndrome. The anemia found in all patients was iron deficient, the mean hemoglobin level was 9.5 g/dl and ferritin rate 10 µg/l. High gastrointestinal endoscopy was performed in all patients; 73 cases (96%) had a single ring while 3 patients had 2 rings (3.9%). Only one patient (1.3%) had 3. All our patients underwent endoscopic dilation with an average of 1.5 dilations with Savary Gillard's bougies in 72 cases (93,5%) and balloons of dilation in 4 cases (5.2%). No cases of perforation were noted after dilatation. The clinical, biological and endoscopic evolution was favorable; no case of malignant degeneration was noted with a mean follow-up of 31.5 months.

Conclusion: Management of PVS is based on endoscopic dilatation and martial supplementation. Our experience confirms that endoscopic dilation is effective, well tolerated and safe.

Regular endoscopic monitoring is required despite endoscopic dilatation because the PVS is a premalignant condition

Keywords: *Endoscopic Dilation; Plummer Vinson's syndrome (PVS)*

Introduction

The first full description of Plummer Vinson's syndrome (PVS) was established by Paterson and Kelly, characterized by the triad: upper dysphagia, iron deficiency anemia, and cervical esophagus fibrous ring (s) [1,2].

Its etiopathogenesis remains poorly defined; the factors most likely to be involved would be iron deficiency, vitamin deficiency and autoimmune origin [3].

The treatment is based on iron supplementation and endoscopic dilation of the esophagus. which is frequently performed to relieve dysphagia due to esophageal stenosis.

Being rare, PVS requires long-term monitoring because it is a pre-cancerous lesion predisposing to the development of squamous cell carcinoma of the esophagus and pharynx [4].

Despite its first description going back more than a century, the PVS literature is limited to isolated clinical cases and small retrospective series. These reports vary widely in terms of management in the absence of clear recommendations. The main objective of our work is to analyze the results of the endoscopic treatment and secondly to study the epidemiological, clinical and endoscopic characteristics of the PVS through a monocentric cohort of 77 patients.

Materials and Methods

This is a descriptive and analytical retrospective study spanning 13 years between September 2005 and June 2018 that included 77 patients collected at the department of functional gastrointestinal exploration of the Ibn Sina University Hospital in Rabat.

All patients in whom the diagnosis of PVS was made based on endoscopy were included: evidence of a post-cricoid oesophageal ring in high endoscopy. All of these patients had dysphagia associated with anemia. All patients with mild esophageal stenosis of another origin or cancer of the upper esophagus were excluded.

Endoscopic dilation of the oesophageal ring was performed in all the patients under general propofol anesthesia under endoscopic vision without fluoroscopic control in most cases by different senior endoscopists. The initial endoscopy made it possible to identify the level of the stenosis from the dental arches, to assess the degree of stenosis, its angulation and its cross-ability or not allowing thus to define the material to be used for the dilation.

Two dilation materials were used:

- The dilation by the Savary-Gilliard bougies (Figure 1).
- Hydrostatic dilatation with balloon (Figure 2).

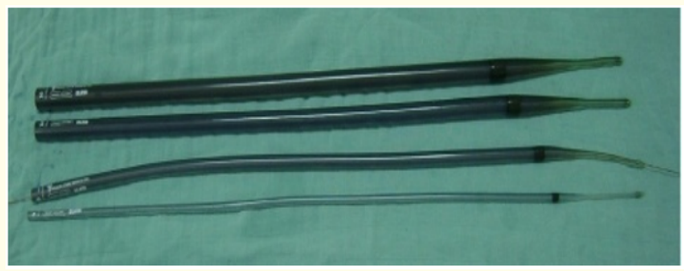

Figure 1*: Savary-Gilliard Bougies.*

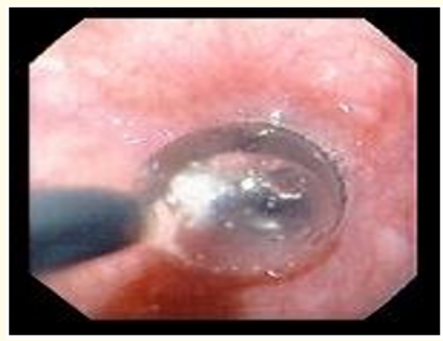

Figure 2*: Dilatation of the PVS ring with hydrostatic balloon.*

Immediate endoscopic control after dilation allowed visualization of the tear in the membrane and study of the rest of the upper digestive tract. Esophageal biopsies were performed to eliminate neoplasia as well as gastric and duodenal biopsies.

The patients were then put under clinical supervision for 6 hours. In the absence of a clinical warning sign, the diet was gradually resumed at night (semi-liquid then normal). The success of the dilatation was defined as an improvement of dysphagia for at least 12 months after the last endoscopic dilatation. A new dilation session was performed in case of recurrence of dysphagia and/or oesophageal stenosis.

Data collection was based on the consultation register, the endoscopy registers of the department and the endoscopic dilation registers. For each patient, we collected the clinical, biological, radiological, endoscopic, anatomopathological, therapeutic and evolutionary data using an exploitation sheet. All the data has been entered on an Excel table and then analyzed by this same software.

Results

We included 77 patients with PVS over a period of 13 years. During this period, 11,373 upper endoscopies were performed, of which 367 allowed the diagnosis of benign organic stenosis of the esophagus, and the PVS accounted for 21% occupying the second position after peptic stenosis. A total of 184 upper endoscopies were performed for the diagnosis and treatment of SPV over 13 years, ie 1.6%.

The average age of the patients was 39 years (range: 16 - 78 years). We noted 2 cases of PVS in adolescents aged 16 and 17 years. There is a female predominance (Figure 3): 66 women (85.7%) vs 11 men (14.3%), with a sex ratio M/F of 1/6. The average patient consultation time was 4 years with extremes ranging from 2 months to 15 years?

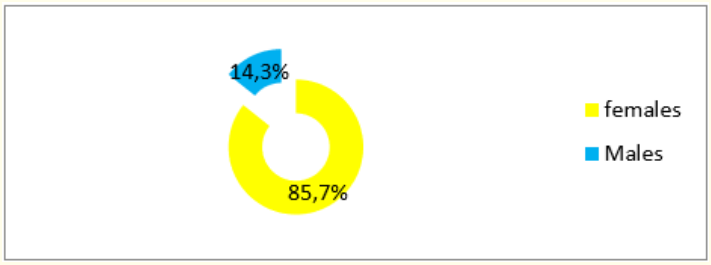

Figure 3*: Distribution of patients with PVS by gender.*

Dysphagia was noted in all patients, progressive worsening in 56 cases (72.7%) and unclear in 21 cases (27.3%) with an average duration of evolution of 4 years. It was permanent in 64 cases (83.1%) and intermittent in 13 cases (16.9%). Its location was high in 74 cases (96.1%) and average in 3 cases (3.9%). 71 cases (92.2%) had selective dysphagia to solids while 6 cases (7.8%) had dysphagia with liquids and semi-liquids.

All patients with PVS had hypochromic microcytic iron deficiency anemia with an average hemoglobin level of 9.5 g/dl (range: 5.5 - 11 g/dl) and mean ferritin of 10 µg/l.

We noted Anemic syndrome in 62 cases (80.5%). 16 cases (20.7%) had skin appendage disorders related to iron deficiency. The considerable and progressive, but not quantified, loss of weight was found in 19 cases (24.7%). Table 1 summarizes the clinical features of patients with SPV in our series.

Symptoms	Number	Percentage (%)
Dysphagia	77	100
Gradual aggravation	56	72,7
Imprecisely	21	27,3
Permanent	64	38,1
Intermittent	13	16,9
High	74	96,1
Average	3	3,9
Solids	71	92,2
Liquids and semi liquids	6	7,8
Anemic syndrome	62	80,5
Weightloss	19	24,7

Table 1: *Clinical manifestations of patients with PVS.*

The upper gastrointestinal endoscopy performed in all our patients showed at least one fibrous ring (Figure 4). 74 cases (96.1%) had an upper esophagus ring; 32 cases (43.2%) in the upper esophageal sphincter and 42 cases (56.8%) in the cervical esophagus. 73 cases (94.8%) had a single ring while three patients had 2 rings (3.9%). Only one patient (1.3%) had 3. During initial diagnostic endoscopy, 3 patients (3.9%) had membrane rupture (Table 2).

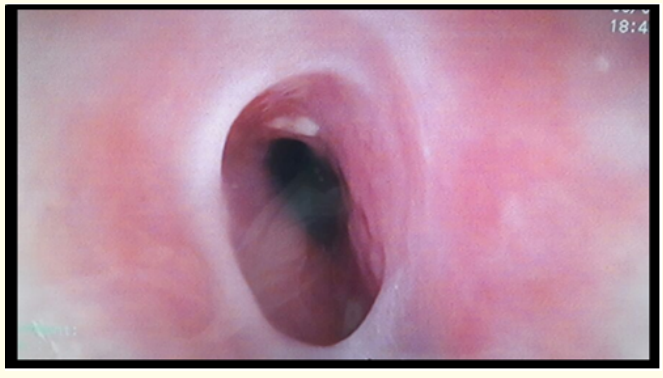

Figure 4*: Endoscopic aspect of the plummer vinson ring.*

Endoscopic aspect	Number	Percentage (%)
Location		
Upper esophagus	74	96,1
Upper esophagal sphincter	32	43,2
Cervical esophagus	42	56,8
Number of rings		
One ring	73	94,8
Tow rings	3	3,9
Three rings	1	1,3
Breaking of the ring	3	3,9

Table 2: *Results of the initial diagnostic upper digestive endoscopy.*

A total of 107 dilation sessions were performed, an average of 1.4 sessions per patient (range: 1 - 5 sessions). It was performed by Savary Gilliard bougies in 72 cases (93, 5%) and dilatation balloons in 4 cases (5, 2%), without fluoroscopic control except in one patient (1.3%) carrying 3 esophageal rings who was dilated by balloon under fluoroscopy.

No case of perforation was noted after dilatation (Figure 5).

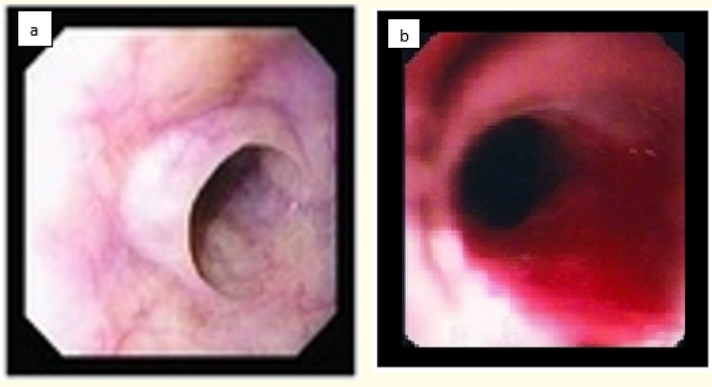

Figure 5*: Endoscopic view before (a) and after (b) dilation.*

In combination with endoscopic treatment, almost all of our patients (n = 75, 97.4%) received systematic oral iron supplementation, only two patients (2.6%) had recourse to a blood transfusion because of clinical intolerance to anemia (Table 3).

	Number	Variation
Dilation by bougies	72	93,5
Dilation by balloon	4	5,2
Iron supplementation	77	100
Per os	75	97,4
Transfusion	210	2,6

Table 3: *Therapeutic aspects.*

The evolution after treatment was favorable in all cases, marked by a disappearance of dysphagia and correction of anemia. The recurrence of dysphagia requiring further dilatation was noted in 11 patients (14.2%). No case of malignant degeneration was noted during patient follow-up with a mean follow-up of 31.5 months.

Discussion

Due to the paucity of international publications and the fragmented nature of the studies, the exact frequency of PVS is not well established. During the first half of the twentieth century, the PVS was particularly common in northern Europe, especially in Scandinavia [5]. The following trend was towards gradual regression due to improved nutritional status [6], therefore short series were published in the literature during this period.

In our department, 77 patients with VPS were collected between 2005 and 2018. On average, the number of patients with VPS per year is between 1.5 and 7.5, the average found in our series is comparable to those found in other series from North Africa [7,8].

The pathogenesis of PVS remains controversial. Many theories have been formulated on this subject, some of which have been abandoned, such as hysteria, streptococcal infection, and tertiary syphilis. Advanced etiopathogenic mechanisms include iron deficiency and nutritional deficits, genetic predisposition, and autoimmune factors. However, the most important theories remain those of primary dysphagia and sideropenia [3,9,10].

The average age of our patients was 39 years, comparable to that of the most recent series [11,12], Jacobs had also found an average age greater than 50 years [14]. The reported epidemiological characteristic of all the studies is the clear predominance of women [7,8,11,12], the sex ratio M/F in our study is lower. Analysis of these data confirms that PVS is prevalent among middle-aged women.

Dysphagia was the most common reason for consultation; it was constantly progressive and painless, which is perfectly concordant with other series [11,13]. Its average duration of evolution was 4 years, comparable to two other series [8,12].

The nemic syndrome comes second and weight loss in the third as functional signs joining the other series [8,12,14]. Weight loss in patients with PVS is quite common; this is due to lack of intake because of the esophageal barrier or an associated disease such as celiac disease. This association has been reported as isolated clinical cases [15,16].

In our series, celiac disease was associated with PVS in 2.6% of cases as in other series [8,14].

PVS is recognized as a pre-cancerous lesion predisposing to hypopharyngeal and esophageal cancer [17], but the development of gastric cancer is less common; only two cases of PVS association with gastric cancer were published in the literature in 2005 and 2008 [10,19]. In our series, the gastric carcinoid tumor was noted in one patient, which is consistent with the literature. Wynder noted that there is a close relationship between SPV and hypopharyngeal cancer [5]. Ahlbom [20] reported cases of two women with SPV for 20 to 30 years who developed respectively five and three different cancers of the lips and oral cavity. No case of association of SPV with hypopharyngeal cancer or tongue cancer has been noted in our series.

Iron deficiency anemia has been found in our series as in other series in most patients with SPV [8,13,21].

In our study as in the literature, the upper gastointestinal endoscopy was performed in all patients allowing us to make the diagnosis of the PVS ring, to specify its number which is in more than 90% of cases unique but, who can be double or triple in rare cases. In our patients, 94.8% had a ring, 3.9% had two rings and 1.3% had 3 rings, whereas in the Fall [12] study, 12% had double stenosis and in the Bakari study [8] 2.3% had a double stenosis. In our series the appearance of the oesophageal ring was annular or semi-lunar interesting the posterior or lateral aspect of the cervical esophagus, this aspect is also found in other works [22].

In our study, as in most other studies [13,16], treatment included routine iron supplementation in combination with endoscopic dilatation. However, in the Niang study [23], 7.5% of patients received exclusive iron supplementation, whereas in other studies, endoscopic treatment was indicated only after martial supplementation [23,24].

Endoscopic instrumental dilation is intended to widen the lumen to remove stenosis-related symptoms, maintain oral nutrition, and prevent inhalation pneumonitis. The mean number of dilation sessions per patient in our work was 1.4 while in the Amouri study [28], it was greater than 2 ± 2.4. The rupture of the ring obtained during the initial endoscopy was noted with small percentages in different series as in ours [8,12,23].

Long-term trends are favorable in most series with low risk of disease recurrence after an average follow-up of 2 years [7,12,23]. In our work, the evolution was positive marked by a disappearance of the dysphagia and a correction of the anemia in all the patients, we did not note recurrence of the PVS nor complications due to the endoscopic dilation.

The formidable complication of SPV is malignant degeneration in squamous cell carcinoma of the esophagus, hence the interest of close endoscopic surveillance. A review of the literature shows that 10% to 30% of cases of VPS evolve to a cancer of the upper digestive tract [11]. In the Lederman series of 106 patients with postcricoid carcinoma, 57% had PVS [25]. In general, 10% of patients with SPV develop pharyngeal or oesophageal cancer [26].

In our work, no case of malignant degeneration was observed during follow-up with a mean follow-up of 31.5 months, this result is comparable to some studies [7,15].

Conclusion

Plummer-Vinson or Paterson-Kelly syndrome is a rare entity that affects mostly adult women. Treatment is based on endoscopic esophageal dilatation and iron supplementation.

Our series is one of the largest series. Our results are comparable to those previously published; our experience confirms that endoscopic dilatation during SPV is safe, effective, well tolerated and relatively easy. However, several sessions are sometimes necessary.

Known as a precancerous condition, regular long-term monitoring should be advocated. In the absence of a recommendation, a high endoscopy is required every year [27].

The prognosis of SPV is excellent, especially since dysphagia and anemia can be effectively treated but become dramatic when combined with squamous cell carcinoma of the hypopharynx or upper esophagus.

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