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Plummer-Vinson syndrome: Clinical, therapeutic and evolutionary aspects: a Moroccan case series

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Abstract: *Plummer Vinson syndrome (PVS) is a rare entity characterized mainly by upper dysphagia, iron-deficiency anemia, and the presence of an esophageal ring on endoscopic examination.*
We conducted a retrospective study over a period of 18 years describing the epidemiological and clinical profile, endoscopic management and the medium and long-term evolution.
A total of 96 patients were enrolled, with a median age of 41,5 years and a clear female predominance (19/77). dysphagia was present in almost all our patients (97,9%), and an anemic syndrome in half the cases (52%). Female gender was significantly associated with PVS occurrence (p=0.002). The frequency of the disease was significantly higher after the age of 30 years with a peak in frequency in the 4th decade (p<0.034).
Upper endoscopy found a membranous ring located in the cervical area in all cases. 1 patient had, in addition to the plummer vinson ring, a lower esophageal membranous called Schatzki ring with a small sliding hiatal hernia. PVS was associated with other autoimmune diseases mostly represented by celiac disease and autoimmune thyroiditis.
Treatment was based on iron supplementation followed by endoscopic dilatation with Savary dilators in 91,6% (n=88) and balloons for 8,3% of cases (n=8). long-term outcome was favorable in 79.1% of patients with no clinical recurrence.
20,8% patients relapsed after a median of 29,2 months [2-72 months] requiring a second dilation.
PVS was complicated by squamous cell carcinoma in 5,2% (n=5).
Keywords: *Dysphagia, Endoscopic dilation, Iron deficiency anemia, Plummer vinson syndrome, Squamous cell carcinoma*

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I. Introduction:

Plummer Vinson syndrome (PVS), also known as Kelly-Paterson syndrome or sideropenic dysphagia, is a rare condition **[1,2,3]**. It is mainly characterized by high dysphagia, iron-deficiency anemia and the presence of an esophageal ring on endoscopic examination, also known as esophageal "web" **[4].** This syndrome mainly affects women, and has been considered a rare entity in Africa since its discovery **[5**]. Treatment is based on esophageal dilatation by dilators or balloon and iron supplementation **[6,7].** However, its identification is essential to determine which patients are at increased risk of developing squamous cell carcinoma of the pharynx and oesophagus **[8]**.

The aim of our study is to describe the epidemiological profile, clinical, biological and evaluate endoscopic features, explore associations with certain autoimmune diseases, and analyze the results of endoscopic dilatation and to compare our results with those of the literature in order to identify the particularities of our population.

II. Materials And Methods:

Study population: This is a retrospective study which collated 96 cases of Plummer Vinson syndrome diagnosed and treated in our hepato gastro enterology C department at Ibn Sina hospital in Rabat over a period of 18 years (from 2005 to 2023).

Were included all patients presenting the triad: dysphagia, iron-deficiency anemia and cervical esophageal web on upper GI endoscopy, and who underwent endoscopic treatment.
patients who have been lost to follow-up were excluded

Data collection: For each patient, we recorded epidemiological, clinical, paraclinical and endoscopic characteristics. Upper GI endoscopy were performed in all patient with systematic gastric, esophageal and jejunal biopsies. Dilatation was performed using a metallic guide wire introduced into the esophageal lumen under endoscopic control without fluoroscopic control. Savary candles are then positioned according to a gradual diameter protocol. The most used dilators were 12 to 15 mm. At the end, a post-dilatation endoscopic check was systematically performed to verify the membranous rupture, the absence of perforation or significant bleeding, and to complete the endoscopic exploration. In the absence of major complications, patients were allowed to resume feeding on the evening of the procedure, starting with a light diet.
Laboratory tests: blood counts and ferritin levels were collected in all our patients.
Statistical analysis: the statistical study was carried out using the jamovi project (2020) (Version 1.6) (Computer Software). Qualitative variables were compared by Khi-2 test and quantitative variables by the student test. All results were considered significant if $p \leq 0.05$

III. Results:

96 patients were collected over a period of 18 years, the median age was 41.5±14.6
[17 to 90 years]. The frequency of the disease was significantly higher after the age of 30 years with a peak in frequency in the 4th decade ($p<0.034$).
A clear female predominance was observed, accounting for 80,2% of cases. (77 female/ 19 male)
Clinically, 4,1% (n=4) patients underwent an antireflux surgery for GERD (gastroesophageal reflux disease) , 3,1% (n=3) patients were active smokers, and 2,08% (n=2) patients had a history of caustic ingestion.
97.9% of patients presented dysphagia as the main symptom, and was permanent in 81% of cases (n=78) associated with a clinical anemia syndrome in 52% (n=50), weight loss in 17.7% (n=17), glossitis in 25% (n=24), cheilitis in 6,25% (n=6), asthenia, splenomegaly,and odynophagia in 3,1% each (n=3).
Biological tests revealed a moderate hypochromic microcytic iron-deficiency anemia in 89.5% of cases(n=86). The mean value of hemoglobin was 8.9 g/dL. A decrease in serum ferritin level without anemia was found in 10% of them (10,4%)

Total of patients	**96**
Median age	41.5±14.6
Sexe ratio (male/female)	19/77
Clinical manifestations :	***(%) n=***
Dyphagia	(97.9%) 95
Anemia	(52%) 50
Glossitis	25% (24)
Splenomegaly	3.1% (3)
Cheilitis	6.25 (6)
Odynophagia	3.1% (3)
Asthenia	3.1% (3)
Biological tests :	***Mean value (range)***
Hb	8.9 (12-18g/dl)
Serum ferritin	16 (25-200 ng/ml)

Table 1 : Baseline characteristics of the study population

A barrium swalow was carried out in 26% of patients (n=25) and revealed a narrowing in the upper esophagus (figure1)

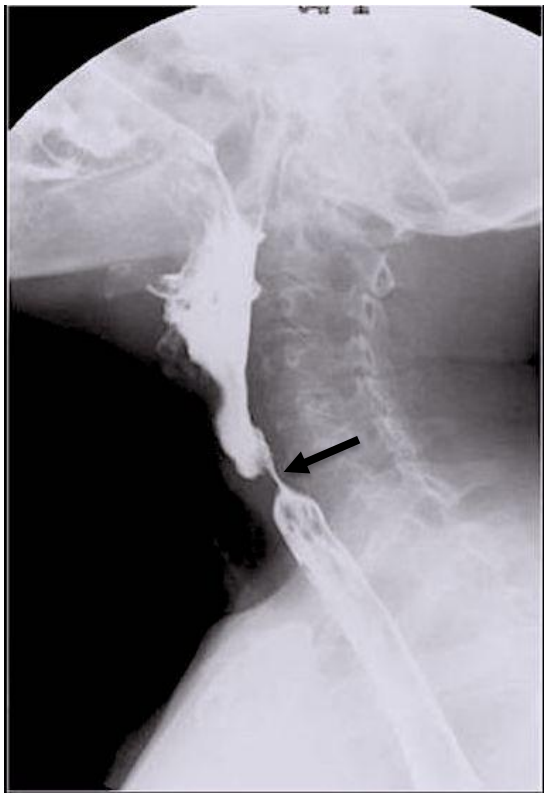
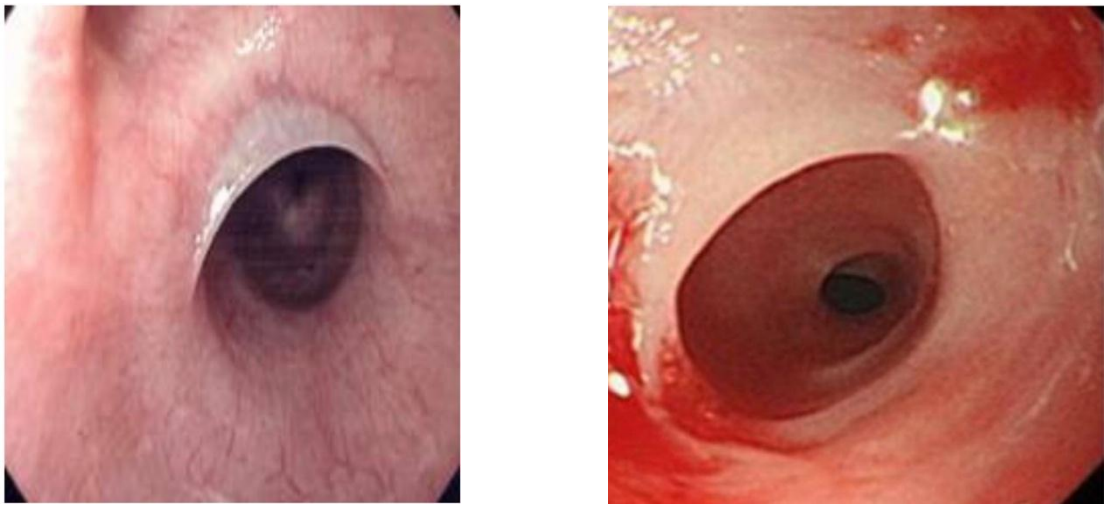


Figure 1 : Barium meal showing tight and short narrowing in the cervical esophagus

All patients underwent oeso-gastro-duodenal fibroscopy, which revealed a membranous ring which is a diaphragm located in the majority of cases just below Killian's mouth (in the cervical area). The ring was semilunar in 26% of cases (n=25) and was concentric in 73.9% (n=71). The ring was impassable in 78% of cases (n=75). dilated spontaneously by the endoscope In 7.2% (n=7) and in 14.5% the membranous was easily crossed (n=14). All patients underwent iron martial supplementation followed by endoscopic dilatation with Savary dilators in 91,6% (n=88) and balloons for 8,3% of cases (n=8), no major pre- or immediate post-procedure complications such as massive bleeding or perforation were recorded.



(a) **(b)**

Figure 2: Endoscopic views of esophageal web thin membranes of esophageal mucosa before (a) and after dilation (b)

Post-dilation control revealed the presence of 2 simultaneous rings (Schatzki ring) with a small sliding hiatal hernia in 1 patient, an associated tumor found concomitantly in another patient whose biopsy showed a

squamous cell carcinoma, a rarefaction of duodenal folds in 2,08% of patients (n=2), and in 2,08% (n=2) patients, the upper endoscopy revealed esophageal varices.
Autoimmune diseases was found represented by celiac disease in 4,1% of cases (n=4), autoimmune thyroiditis in 4,1% (n=4), crohn's disease in 2,08% (n=2), rhumatoid arthritis in 2,08% (n=2) and autoimmune bullous dermatosis in 1 patient. Portal hypertension was found in 3.1% of our patients (n=3).

Associated autoimmune diseases	Number of patients
Celiac disease	4
Autoimmune thyroiditis	4
Crohn disease	2
Autoimmune bullous dermatosis	1
Rhumatoid arthritis	2

Table 2 : Associated autoimmune diseases in plummer-vinson syndrome

Long-term outcome was favorable in 79.1% of patients with no clinical recurrence, with a median follow-up was 9 months
20,8% patients (n=20) relapsed after a median of clinical remission of 29,2 months [2-72 months] requiring a second dilation.

During follow-up, 4,1% of patients developed squamous cell carcinoma (n=4), and in 1 patient, squamous cell carcinoma was found at the time of diagnosis. (figure 4)
Female gender was significantly associated with PVS occurrence (p=0.002), while the duration of dysphagia and history of reflux surgery was not statistically significant (p=0.841)

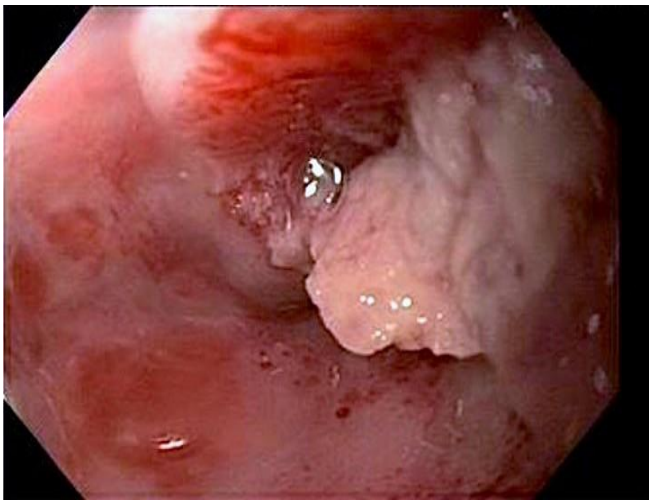


Figure 3 : Squamous cell cancer of the proximal oesophagus

IV. Discussion:

Until the 60's, PVS was most often described in the Caucasian population, mainly in Scandinavian and Northern European countries. **[14,15]** Today, it is a very rare entity worldwide, especially in Africa.**[14,16]**
the aim of our case study was firstly to determine the epidemiological, clinical and therapeutic features of PVS, and then to identify the particularities of our population by analyzing the therapeutic and evolutionary modalities and comparing them with the literature.
PVS is rare. PVS is uncommon. Most literature points to a female predominance, accounting for 90% of cases. It is also more occur more frequently in people aged between 40 and 70, although it has been recently noted in young children (<6years old). **[12,13]**
In our study, a clear female predominance was observed, accounting for 80,2% of cases and the frequency of the disease was significantly higher after the age of 30 years with a peak in frequency in the 4th decade.

Clinically, It typically presents as progressively increasing cervical dysphagia, initially involving solid foods in iron-deficient women **[17]**.
Iron-deficiency anemia is very common and is suspected on the basis of a clinical anemia syndrome. The most frequent symptoms of anemic syndrome are phyisical asthenia, tachycardia, cutaneous-mucosal pallor and dyspnea. Other symptoms include alopecia, dry skin and hair, glossitis, koilonychia, vertigo and syncope.
In our study, clinical signs other than dysphagia and anemia were glossitis, cheilitis, odynophagia and splenomegaly.
Biologically,Laboratory evaluation is necessary to etablish iron deficiency cause. Tests include : Hemoglobin, the mean cell volume (MCV) , mean cell hemoglobin (MCH) and serum iron studies (serum iron, ferritin and total iron binding capacity) who can be decreased in chronic anemia.
Upper endoscopy is the reference test for all dysphagia. This examination has the advantage of allowing both diagnostic and therapeutic measures. However, this procedure is still perceived as being less effective than the barium contrast study in detecting esophageal rings. **[20]**. The reason is, Esophageal webs are very thin and close to the upper esophageal sphincter, making them difficult to detect. They can also be dilated by the endoscope and ruptured without being seen. In the case of any suspected oesophageal ring or oropharyngeal dysphagia, the endoscopist must refine the exploration to identify any anomalies, mainly in the cervical region . **[21]**
The majority of esophageal webs are single and either circumferential or anterior in position, but multiple webs or posterior in location may also occur **[19].**
Sometimes,Schatzki rings can also be found on the lower esophageal area. Most are asymptomatic but are considered to be the most common cause of episodic dysphagia in adults. Usually it is associated with other esophageal disorders such as gastroesophageal reflux and which may actually be causally related to it **[18].** in our study, one patient had the combination of 2 esophageal rings

Therapeutic management of PVS consists firstly in correcting iron deficiency through supplementation, usually by injection, after eliminating any other cause of iron deficiency anemia, such as celiac disease, digestive hemorrhage or tumor. **[1,2,22]** Secondly, treatment is based on endoscopic dilatation, usually using Savary-Gilliard dilators, or balloon dilatation. **[1,2,23]**
The treatment for esophageal ring can be carried using Savary Gilliard dilators, the technique involvinf the introduction of a metallic guidewire through the oesophageal web under endoscopy and into the stomach. The endoscope is then removed and dilators of increasing size are progressively introduced, generally from 12 to 15 mm in diameter 24.
Independently of the therapeutic aspect, endoscopy can also be used to detect and frequently monitor other potentially associated malignancies, such as post-cricoid, oral, pharyngeal, esophageal, stomach and colon malignancies, the incidence of which can be as high as 16% in patients with PVS. **[24,25,26]**
In other words, in our study, 5 patients presented a degenerescence of the plummer vinson web.
Generally, patients with post-cricoid web or PVS have an excellent outcome. PVS is considered to be a precancerous state
There are no formal guidelines for the follow-up of patients with PVS. Considering the malignant potential, annual endoscopic surveillance is recommended by most experts, depending on risk factors and clinical circumstances. **[9,10,11]**

V. Conclusion:

PVS is relatively rare and mainly affects women. Although endoscopic dilatation has proved effective in most cases, in our study, recurrence involved 20% of patients. Regular follow-up is important because of the risk of degeneration and the high frequency of associated autoimmune diseases. However, correction of iron-deficiency anemia remains crucial to prevent disease recurrence.

Data availability statement
The raw data supporting the conclusions of this article will be made available by the authors without undue reservation.

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Conflict of Interests : The authors declare that there is no conflict of interest.

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Figure legend:

Table 1: Demographic and clinical characteristics of the study population

Table 2: Associated autoimmune diseases in plummer-vinson syndrome

Figure 1: Barium meal showing tight and short narrowing in the cervical esophagus

Figure 2: Endoscopic views of esophageal web thin membranes of esophageal mucosa before (a) and after dilation (b)

Figure 3: Squamous cell cancer of the proximal oesophagus