Pemphigus and Plummer-Vinson syndrome: an uncommon association

ABSTRACT

Aims : The reported association of Plummer-Vinson syndrome and some autoimmune diseases, including pemphigus -association described for the first time-, raises the hypothesis of the autoimmune origin of this syndrome. On the other hand, this unexpected association represents a therapeutic challenge given the risk of the progression of pemphigus lesions after an esophageal dilatation session, hence the originality and interest of our case.

Case report : A 60-year-old patient was admitted to our department for pemphigus foliaceus and treated with oral corticosteroids. The workup revealed an iron deficiency anemia. A dysphagia to solids evolving for 4 years led to an esophageal endoscopy which revealed the presence of esophageal ring. The diagnosis of Plummer-Vinson syndrome was retained but the dilatation could only be done after stabilizing the pemphigus to avoid a detachment of the esophageal mucosa by Koebner phenomenon.

Discussion : Plummer-Vinson syndrome is a rare condition defined by the presence of dysphagia, iron deficiency anemia and esophageal ring. Its etiopathogeny is poorly understood. It can be associated with some autoimmune pathologies suggesting an autoimmune origin. To date, no association with pemphigus has been described in the literature.

Conclusion : The autoimmune origin of Plummer Vinson syndrome is to be discussed in regard to the described associations with autoimmune diseases which would not be random.

Keywords: Plummer-Vinson - Dysphagia - Anemia - Esophageal web - Pemphigus - Autoimmune disease

1. INTRODUCTION

Plummer-Vinson syndrome (PVS), also known as Kelly-Patterson syndrome, is characterized by a classic triad of dysphagia, iron deficiency anemia and esophageal webs. It is commonly considered as a precancerous condition. Its potential association with autoimmune diseases is acknowledged, but to date no association with pemphigus has been reported. We thus report the first case of pemphigus associated with a Plummer-Vinson syndrome.

2. PRESENTATION OF THE CASE

A 60-year-old male patient, with no medical history, was admitted to the dermatology department for a bullous dermatosis with slight pruritus, predominantly in seborrheic areas, on normal-appearing skin, evolving for one year with a negative Nikolsky's sign. The involved skin surface was estimated at 60% with a Pemphigus Disease Area Index (PDAI) of 36 (Figure 1a,b). Mucous membranes were spared. A skin biopsy was performed with histopathological study and direct immunofluorescence as well as an anti-intercellular substance antibodies testing (Figure 2,3). All results were indicative of pemphigus foliaceous. An additional workup was performed to detect any underlying associated disorders. It only showed a mild iron deficiency anemia with a hemoglobin level of 11 g/dl.
Oral corticosteroid-therapy, associated with classic adjuvant measures, was initiated at a dose of 2mg per kg a day. Intravenous iron supplementation was introduced as well.
On the other hand, our patient suffered from an upper dysphagia to solids, evolving for about four years, which required an endoscopic management. It revealed the existence of an esophageal web at 22 centimeters from the dental arches (Figure 4).The diagnosis of Plummer-Vinson syndrome was then made. The patient was to undergo esophageal dilatation using a Savary-Gilliard dilator, but this could only be done after stabilization of the pemphigus and complete clearance of the skin lesions, which took more than nine weeks of oral corticosteroid-therapy.
Follow-up showed a total resolution of dysphagia after dilatation, as well as complete recovery of pemphigus skin lesions after 16 weeks of corticosteroid therapy. After a 4-year follow-up, there was no recurrence of either pemphigus or Plummer-Vinson syndrome.

3. DISCUSSION

Plummer Vinson syndrome is a rare condition that mainly occurs in caucasian middle-aged women [1]. The incidence of the syndrome is declining, which may be ascribed to better nutrition and iron deficiency treatment [2]. Considered as a precancerous condition, malignant transformation, into squamous cell carcinoma in the majority of cases, occurs in 3 to 15% of all patients. Therefore, it requires regular and close monitoring [3,4]. Its etiopathogenesis remains poorly understood. The most significant possible risk factor is iron deficiency in people with genetic predisposition [5,6]. Other includes vitamin deficiencies such as riboflavin, pyridoxine and thiamine [7,8]. Nevertheless, the autoimmune hypothesis remains relevant, given the possible association of PVS with different autoimmune diseases, which according to some authors, is not a coincidence [9].

In fact, PVS can be associated with Biermer's disease, coeliac disease, Crohn's disease, rheumatoid arthritis, thyroiditis or Goujerot-Sjögren's syndrome [5,10,11,12,13]. To our knowledge, no association with pemphigus has been reported before our case. However, further reports are needed to clarify whether the coexistence of PVs and pemphigus is casual or not.

Management of PVS is mainly based on iron supplementation and endoscopic esophageal dilatation using special dilators [4]. In the case of our patient, this procedure was not devoid of risk. As pemphigus is an autoimmune bullous dermatosis with an increased risk of Koebner's phenomenon, dilatation could induce an extensive sloughing of the esophageal mucosa. We were then compelled to delay the dilatation procedure until stabilization of the pemphigus under high-dose corticosteroid-therapy. The outcome was successful with complete resolution of the dysphagia and clearing of the pemphigus lesions.

4. CONCLUSION

Despite the prevalence of iron deficiency anemia, Plummer-Vinson syndrome remains a rare and underreported condition worldwide. Its association with autoimmune diseases is more and more often observed and raises many doubts as to the possible autoimmune origin of this syndrome. In our case, the association with pemphigus was not only unprecedented, but also represented a real therapeutic challenge given the significant risk of Koebner's phenomenon induced by esophageal dilatations.

CONSENT

All authors declare that written informed consent was obtained from the patient (for publication of this case report and accompanying images).

ETHICAL APPROVAL

All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

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FIGURES

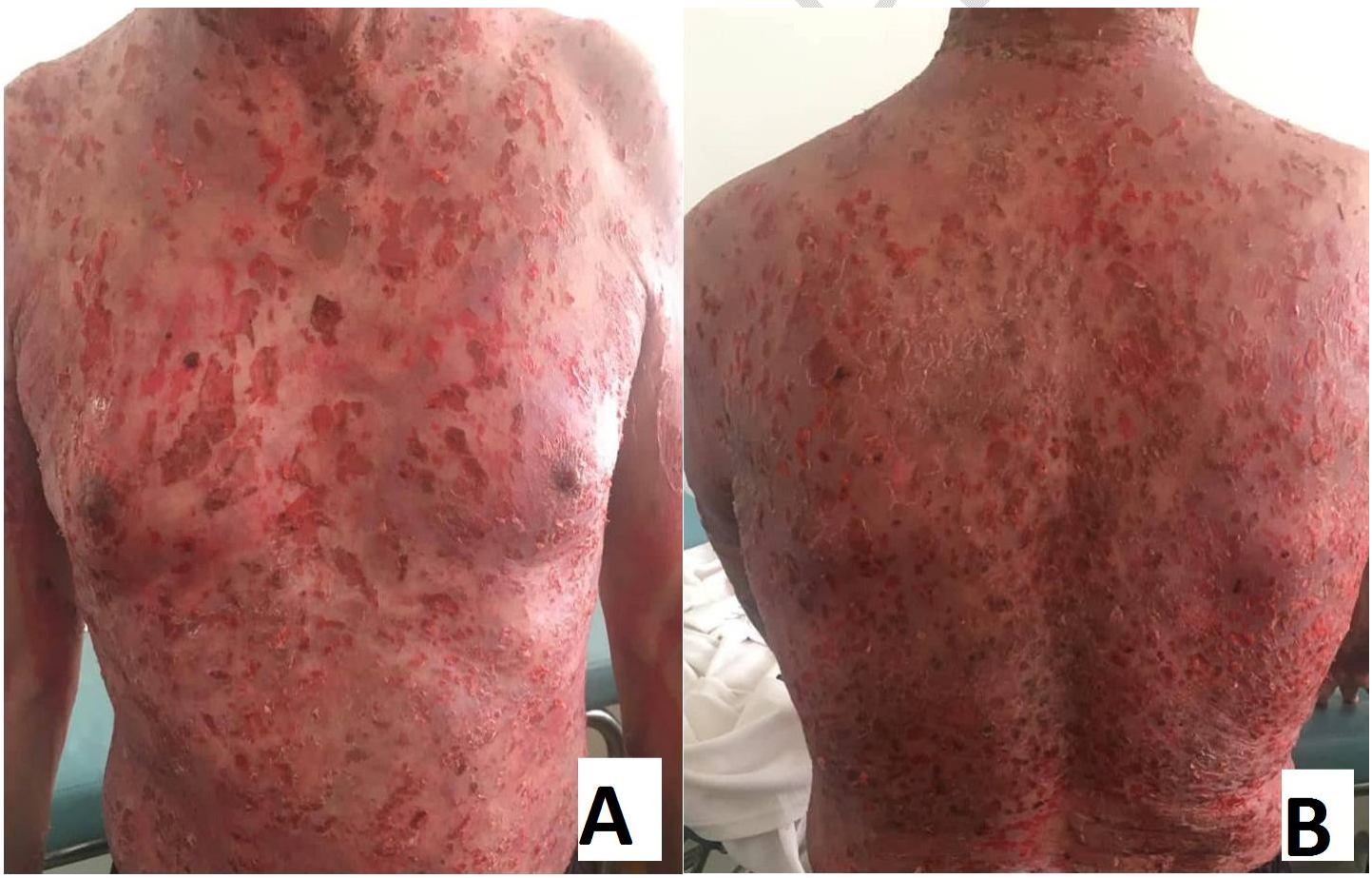

Figure 1 : Multiple post-bullous erosions on healthy skin covering the entire trunk.

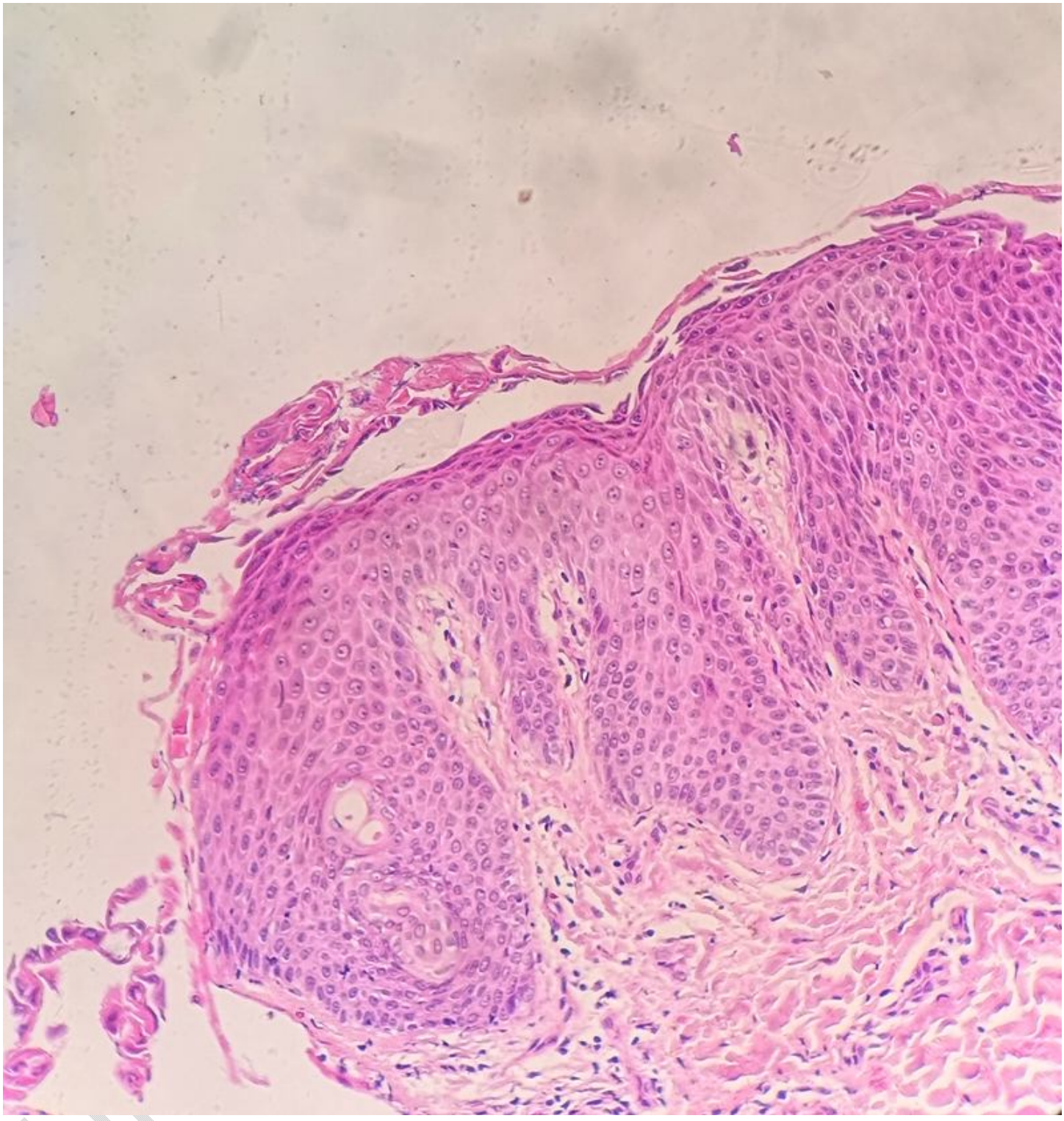

Figure 2 : Histological section of a post-bullous erosion biopsy showing superficial intraepidermal cleavage with acantholytic cells (Hematoxylin and eosin).

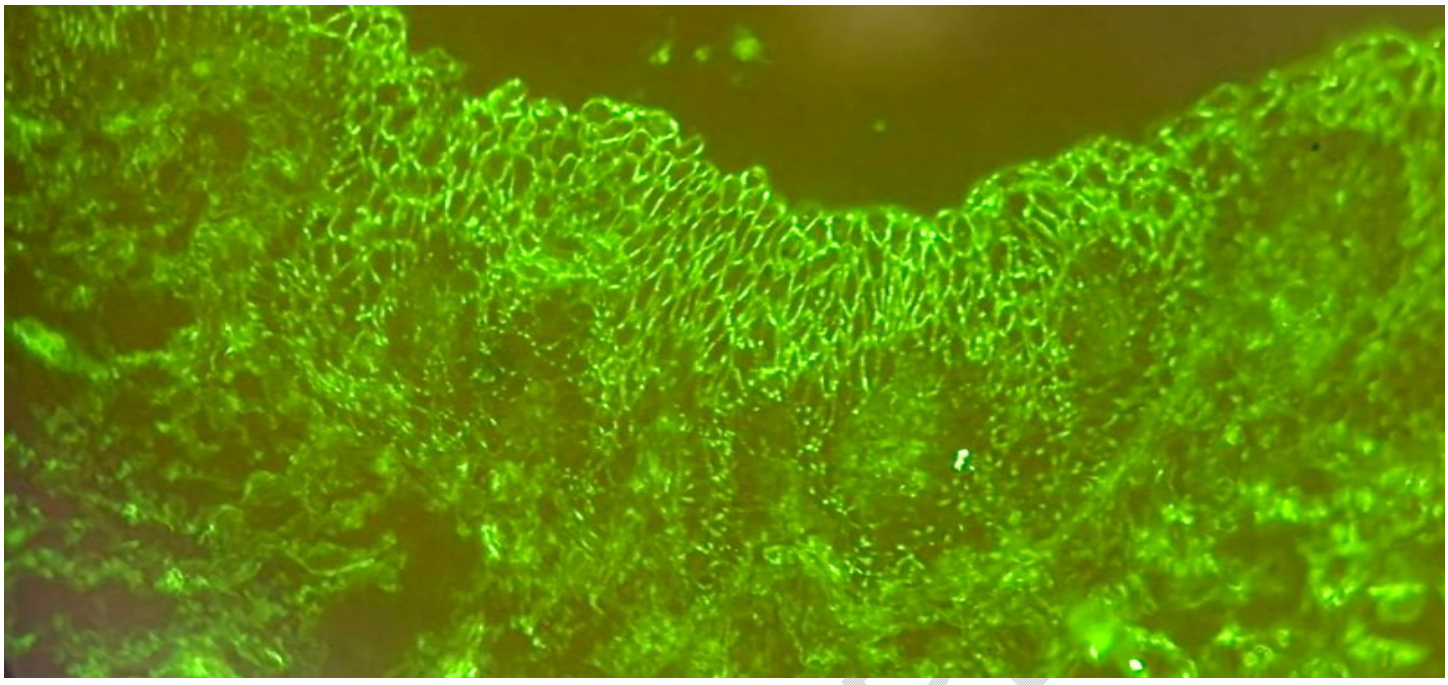

Figure 3 : Direct immunofluorescence on healthy skin showing a gridded deposit of IgG in the epidermis.

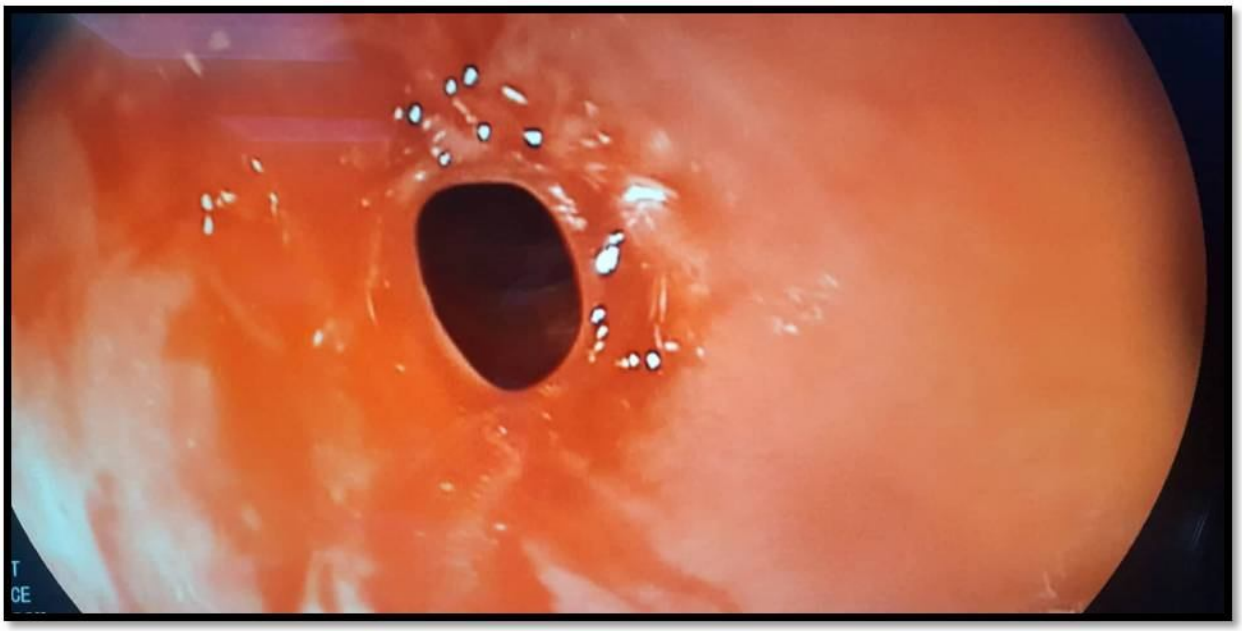

Figure 4 : Endoscopic image showing an esophageal ring located 22cm from the dental arch.