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PLUMMER-VINSON SYNDROME IN A PATIENT WITH NECROBIOSIS LIPOIDICA: REPORT OF AN UNUSUAL ASSOCIATION

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ABSTRACT

Plummer-Vinson Syndrome (PVS), also known as Kelly-Paterson Syndrome, is a rare condition that usually affects middle-aged white women between 40 and 70 years old, being very uncommon in males and presenting itself through a classic triad, consisting of dysphagia for solids, iron deficiency anemia and esophageal membrane. Necrobiosis lipoidica (NL) is a rare, degenerative connective tissue disease that presents with palisade-like granulomatous skin inflammation. It occurs mainly in young adults, being three times more in women. The present report demonstrates a case of a 49-year-old woman, with necrobiosis lipoidica, admitted to the gastroenterology outpatient clinic of the HELGJ (Hospital Escola Luiz Gioseffi Jannuzzi, Brazil) being investigated and diagnosed with PVS, fulfilling the three classic requirements of the syndrome (dysphagia, anemia and esophageal membrane), the two diagnoses that, despite being rare, have no correlation, and their incidence in the same patient is quite uncommon.

KEYWORDS: Esophageal membrane; iron deficiency anemia, dysphagia.

INTRODUCTION

Plummer-Vinson syndrome (PVS), also known as Kelly-Paterson syndrome, is a rare condition that usually affects middle-aged white women between 40 and 70 years old, being very uncommon in males and presenting itself through a classic triad, consisting of dysphagia for solids, iron deficiency anemia and esophageal membrane.[1] Etiology is still not well defined, with theories that suggest for autoimmunity, genetic predisposition, malnutrition, among others. In the histopathological analysis, microscopy shows epithelial atrophy with chronic inflammation of the submucosa and epithelial atypia, which may present dysplasia in some more advanced cases.[1,2] Chronic irritation of the esophageal mucosa predisposes the formation of esophageal membranes and strictures, with hypopharyngeal, esophageal and oral neoplasms as one of the complications of the disease. In the physical examination of these patients, the presence of cutaneous-mucous pallor due to anemia, malnutrition due to dysphagia and esophageal stenosis, as well as weakness, adynamia, fatigue and glossitis are observed.[2] In the complementary exams, the presence of anemia is confirmed. iron deficiency, and the barium esophagogram is the best test initially used to evaluate esophageal stenosis and the presence of esophageal membranes, which can be better observed by upper digestive endoscopy (UDE), an exam which allows the therapeutic action of esophageal dilation and in in some cases, correcting dysphagia. Iron deficiency anemia is multifactorial, being precipitated by dysphagia itself, which impairs the nutritional status of these patients, as well as an increase in inflammatory mediators such as procalcitonins, being treated only with iron replacement.[2,3]

Although Necrobiosis lipoidica (NL) is a rare, degenerative connective tissue disease that presents with palisade-like granulomatous skin inflammation. It occurs mainly in young adults, being three times more in women. Lesions present as erythematous papules or plaques that grow centrifugally and turn yellowish-brown, with atrophy and telangiectasias in the center and raised erythematous borders. They can be single or multiple and often coalesce.[4] In most cases, the lesions are bilateral and occur on the lower limbs, with the

pretibial region being affected, but lesions on the scalp, face, trunk, penis or even diffuse lesions may occur. Lesions tend to be chronic, with variable progression, and there may be long periods of quiescence or resolution.[5] There is an association of NL with diabetic patients (DP), and sixty-five percent of patients with NL also have diabetes mellitus (DM); however, the disease occurs in only 0.3% of the DM population.[6] The diagnosis of LN is confirmed through histopathology, whose most striking feature is the inflammatory process involving the subcutaneous tissue and the coexistence of similar lesions in the dermis, with horizontal bands of inflammatory cells, interspersed with areas of degenerated collagen and fibrosis, involving the entire dermis thickness. The presence of an inflammatory infiltrate composed predominantly of neutrophils in the septum is characteristic of early stages of the disease, with follicular lymphoid formation being observed in the thickened septum of old lesions.[4,5] When the lesions become atrophic, the inflammatory infiltrate decreases and small granulomas remain with multinucleated giant cells in the midst of fibrotic and degenerated collagen. In addition to the aesthetic alteration and the painful condition caused by the lesions, they are subject to secondary infections, and therefore the treatment of the disease is important.[6] There are several treatment modalities described, but all with limited success: corticosteroids - topical, intralesional and systemic -, topical retinoids, topical tacrolimus, antiplatelet drugs such as aspirin or ticlopidine, agents that lower blood viscosity such as pentoxifylline, mycophenolate mofetil, fumaric acids, cyclosporine, chloroquine, thalidomide, PUVA, photodynamic therapy and anti-TNF drugs.[4,5]

CASE REPORT

Female, 49 years old, with asthma, chronic ulcerative lesions in the lower limbs for 30 years, biopsy-confirmed necrobiosis lipoidica, uterine cancer (squamous cell carcinoma IIb) after 35 radiotherapy sessions started in 2018. She denies systemic arterial hypertension, diabetes, drug allergies, alcoholism and reports passive smoking for 5 years. In home use of torsilax, alenia and omeprazole, but reports previous use of corticosteroids for a long period due to skin lesions. She comes to the gastroenterology outpatient clinic complaining of epigastric pain that started after radiotherapy 2 years ago. He also reports episodes of melena and bright blood in the stool, coursing with sporadic episodes of liquid diarrhea, without mucus or pus. In addition, she reports dysphagia for solid foods for 8 months. She had pale mucous membranes and scaly and hyperchromic lesions up to the thigh root, extensive ulcerations up to the metatarsal region and significant nail atrophy. showing no other relevant changes on physical examination. Laboratory examination showed mild anemia, with hemoglobin of 10.3g/dL. In view of the case, upper digestive endoscopy was requested, which showed stenosis of the proximal and middle esophagus compatible with the presence of esophageal membranes compatible with Plummer-Vinson Syndrome, requiring a new upper digestive endoscopy for esophageal dilation with Savary-Gilliard candles. Since there were no bleeding lesions that could justify the melena and hematochezia, a colonoscopy was performed, which showed hemorrhoidal disease. In a second moment, endoscopic esophageal dilation was performed, without intercurrences, with usual medication and proton pump blocker, maintaining outpatient follow-up in the gastroenterology service.

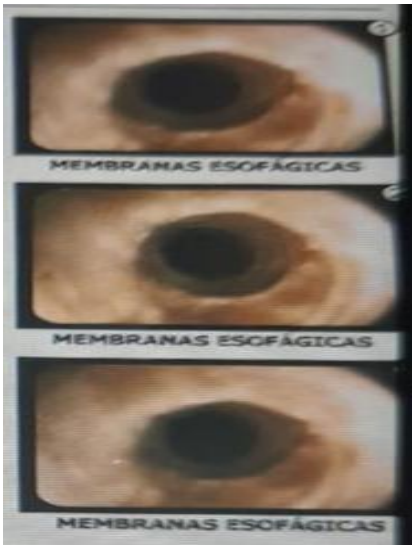

Image 1- Esophageal membranes at upper digestive endoscopy.

DISCUSSION

The patient in question went to the Gastroenterology outpatient clinic of Hospital Escola Luiz Gioseffi Jannuzzi in Valença – RJ, Brazil to clarify epigastralgia, dysphagia, melena and hematochezia. In addition, he is being monitored for two rare syndromes which he has: Plummer-Vinson Syndrome (PVS) and Lipoid Necrobiosis. Although they manifested concomitantly in this patient, they do not have any type of clinical relationship.

PVS has a classic triad, consisting of dysphagia for solids, iron deficiency anemia and the presence of esophageal membranes.[1] Possible risk factors include malnutrition, genetic predisposition and autoimmune diseases.[2] In addition to the complaint, laboratory alterations and epidemiological range being compatible, the patient in The question was submitted to an upper digestive endoscopy (UDE) which showed stenosis of the proximal esophagus and the presence of esophageal membranes. The other component of the triad, iron deficiency anemia, is evidenced by the presence of hemoglobin 10.3 mg/dl.2,3 Once the documented classic triad and the usual epidemiological age group are present, the diagnosis of Plummer Syndrome can be confirmed. -Vinson. Other causes of dysphagia besides PVS were ruled out, as they are more common.

In turn, necrobiosis lipoidica, as previously mentioned, is considered a rare and connective disease of the connective tissue, presenting with granulomatous cutaneous inflammation.[4,5] Some authors suggest that NL is an immune complex vasculitis with other factors such as abnormal production of collagen, diabetic microangiopathy or altered neutrophil migration.[6] Although a relationship between diabetes and NL has already been demonstrated, the patient in this report is not diabetic. The patient was diagnosed 27 years ago with NL and having a 1993 biopsy compatible with the disease itself, this being the other standard exam for diagnosis.[5] Since then, she has had relapses and remissions of the disease, requiring some hospitalizations for treatment.

CONCLUSION

Although the patient in the case presented here has both Plummer-Vinson Syndrome and Necrobiosis lipoidica, these, in turn, do not have any clinical relationship, occurring independently of each other, and their incidence in the same patient is quite uncommon. It is important to note that there are other more common differential diagnoses for the aforementioned signs and symptoms.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

CONSENT

All authors declare that 'written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office/Chief Editor/Editorial Board members of this journal.

ETHICAL APPROVAL

Not applicable.

BIBLIOGRAPHIC REFERENCES

1. Field, Zachary et al. Plummer-Vinson syndrome and heart failure: an unusual association in an African American woman. The American journal of case reports, 2019; 20: 1264.
2. KARTHIKEYAN, Priyadarshini; ASWATH, Nalini; KUMARESAN, Ramesh. Plummer Vinson syndrome: a rare syndrome in male with review of the literature. Case reports in dentistry, v. 2017, 2017.
3. GADE, Ajay K.; PACHECO, Lauren. A rare case of plummer-vinson syndrome. Cureus, 2019; 11: 12.
4. PEYRÍ, Jordi; MORENO, Abelardo; MARCOVAL, Joaquin. Necrobiosis lipoidica. In: Seminars in cutaneous medicine and surgery, 2007; 87-89.
5. TOKURA, Yoshiki et al. Necrobiosis lipoidica of the glans penis. Journal of the American Academy of Dermatology, 2003; 49(5): 921-924.
6. O'TOOLE, E. A. et al. Necrobiosis lipoidica: only a minority of patients have diabetes mellitus. The British journal of dermatology, 1999; 140(2): 283-286.