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Celiac disease and IgA vasculitis; association celiac disease and rheumatoid purpura: A case report and a review of the literature

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Article History Received: 25.04.2020 Accepted: 22.05.2020 Published: 29.05.2020 **Abstract:** Celiac disease is an autoimmune enteropathy that can be associated with other autoimmune diseases including igA vasculitis: rheumatoid purpura. We report the case of a patient with celiac disease on a gluten-free diet who developed a rheumatoid purpura clinically suspect and confirmed by anatomopathological study of skin biopsies. Rheumatoid purpura in adults is a rare entity but possible as an association with celiac disease, the pathophysiology involves igA, the transferrin and transglutaminase receptors, the prognosis depends on the renal and digestive system, the treatment is often symptomatic.

Keywords: celiac disease, autoimmunity, igA vasculitis, rheumatoid purpura.

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CASE REPORT

INTRODUCTION

Celiac disease is a chronic autoimmune enteropathy occurring in genetically predisposed HLA-DQ2 or DQ8 subjects. It can be associated with other autoimmune diseases such as autoimmune hepatitis, thyroiditis and IgA nephropathies.

Our case describes the possible association of celiac disease and igA vasculitis such as rheumatoid purpura.

CASE REPORT

We report the case of a 43-year-old man, followed for 2 months for celiac disease retained in front of positive igA antitransglutaminase AC and subtotal villous atrophy of stage 3b according to the modified Marsh classification, on a gluten-free diet complicated by exudative enteropathy with polyseritis made of bilateral edema of the lower limbs, ascites and bilateral hydrocele, admitted for a purpuric and ecchymotic spots extensive involving the 2 lower limbs (Figure 1), associated with acute symmetrical abdominal pain and bilateral, inflammatory arthralgia of the lower limbs involving

the knees and ankles, evolving in a context of febrile sensation and profound deterioration in the general condition, an abdominal ultrasound is performed to eliminate a surgical emergency income normal, positive inflammatory with CRP at 300 and white blood cells at 8010 with lymphopenia at 1030, normal platelet count at 365,000, hypoalbuminemia at 14.8 and 24-hour proteinuria negative, blood cultures negative, after eliminating the hematological and infectious origin of purpura, a skin biopsy performed was in favor of vasculitis with IgA deposits.

The treatment consisted of a strict rest, corticotherapy 1mg / kg / d cover by antibiotics based on C3G with an albumin infusion; the evolution was marked by a good clinical and biological improvement from Day 3 of corticotherapy with regression of purpuric lesions, disappearance of abdominal pain and regression of the intensity of arthralgia.



Figure-1. Purpuric and bruising spots on the thigh roots

Discussion

Celiac disease is often accompanied by autoimmune disease, which tops the list of type I diabetes, autoimmune thyroiditis, autoimmune hepatitis and biermer disease (Nion-Larmurier, I. & Cosnes, J., 2009).

The association celiac disease and igA vasculitis has been described with Berger's disease (igA nephropathy) with implications of igA, the transferrin and transglutaminase receptors, thus the interest of the gluten-free diet in the disappearance of igA deposits and improvement of the condition of the glomeruli (Papista, C et al., 2015). The same mechanism can explain the occurrence of rheumatoid purpura during celiac disease with deposits of igA on small vessels.

Rheumatoid purpura is rare in adults in the order of 1 in 1 million, with a male prevalence (Watts, R. A *et al.*, 2005). The short-term prognosis depends on the severity of the digestive and long-term renal involvement. Pillebout, E. & Verine, J. 2014).

Digestive manifestations are frequent, variable depending on the series; these are spasmodic pain, moderate but can be severe leading to laparotomy. They can be associated with a digestive hemorrhage, at Endoscopy, an aspect of petechial purpura or even real necrosis areas of the digestive wall. More rarely, other organs such as the lung, heart or nervous system may be affected. Skin (leukocytoclastic vasculitis) and renal (proliferative endocapillary glomerulonephritis) histology, associated with the presence of IgA deposits in these tissues, confirm the clinical diagnosis. Pillebout, E. & Verine, J. (2014).

Recent publications in the pediatric and adult series thus show the existence of chronic renal failure, which progresses sometimes more than ten years after the first attack (Pillebout, E. *et al.*, 2002; Coppo, R. *et al.*, 1997). The treatment of rheumatoid purpura is most often symptomatic. The benefit of

more specific treatments (corticosteroids or immunosuppressants) during severe visceral forms (most often abdominal or renal) has not yet been established (Huber, A. M. *et al.*, 2004; Foster, B. J. *et al.*, 2000).

CONCLUSION

Several autoimmune pathologies can be associated with or indicative of celiac disease, notably rheumatoid purpura rarely described in adults and exceptionally associated with celiac disease, the pathophysiology of which is attributed to the involvement of igA in both pathologies.

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