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Clinical Image

Necrosis of the nose revealing chronic agglutinin disease

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Chronic agglutinin disease (CAD) is an extraordinary shape of grownup autoimmune hemolytic anemia because of the presence of an IgM autoantibody this is energetic at low temperature and leads to the multiplied destruction of pink blood cells for the duration of supplement activation.

The management of CAD, which has long been empirical and mostly symptomatic, has improved with the use of rituximab alone or combined with chemotherapy in order to target the underlying B-cell clone. Moreover, the ongoing development of new complement inhibitors opens a promising era for the management of patients with severe and transfusion-dependent hemolysis [1].

We report the case of a 67-year-old patient with type 2 diabetes with degenerative complications treated by insulin analogues who consulted for dry necrosis of the nose (Fig. 1) evolving for a week in a context of apyrexia with weight loss of 5 kg in 2 months.

On biology there was no autoimmune hemolytic anemia. The antiphospholipid syndrome assessment and complement dosage were normal. Tumor markers were negative. The blood count showed an agglutination of red blood cells at a temperature below 37°C. The diagnosis of chronic agglutinin disease was made and an etiological investigation was started to search a secondary origin, in particular the Waldenstrom disease.

Differential diagnoses

1. Raynaud's syndrome
2. Vasculitis

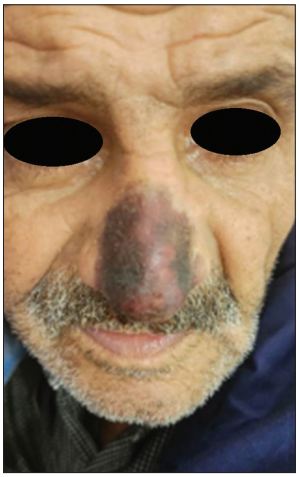

Figure1: Dry necrosis of the nose in patient with chronic agglutinin disease.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

REFERENCE

1. Lamarque M, Michel M. La maladie des agglutinines froides: spectre clinicobiologique et données d'une étude rétrospective multicentrique française portant sur soixante-neuf patients. Hématologie. 2017;23:366-78.

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