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LETTER TO THE EDITOR

Atypical presentation of anti-NXP-2 positive juvenile dermatomyositis

Manifestação atípica de dermatomiosite juvenile com anti-NXP-2 positivo

*Andreia Coimbra-Sousa, Renata Miossi, Fernando H. C. de Souza, and Samuel Katsuyuki-Shinjo[a]**

Division of Rheumatology, Faculdade de Medicina FMUSP, Universidade de São Paulo, São Paulo, Brazil
[a]ORCID: 0000-0002-3682-4517

Dear Editor,

The anti-NXP-2 is one of a myositis-specific autoantibody considered a marker of dermatomyositis (DM)[1,2]. In addition, there is a strong relationship between NXP-2 autoantibodies and calcinosis, particularly in juvenile DM[2]. In this context, we reported an atypical presentation and evaluation of a patient with anti-NXP-2 positive juvenile DM.

A female patient presented with whitish papules on her right forearm at 8 years of age. Seven years thereafter, the patient had proximal muscle weakness of the upper and lower limbs associated with transmission dysphagia, elevated CPK (maximum 350U/L), magnetic resonance imaging of the thighs with evidence of muscle edema, and a non-specific muscle biopsy. There was calcinosis over the previous lesion on the right forearm (Fig. 1A) as well as on other pressure points (such as armpits and posterior face of the knees). She denied any cutaneous changes, including heliotrope rash, Gottron's papules, and sign. A skin biopsy of the whitish papules revealed mild perivascular and perianexal dermatitis with foci of vacuolar alterations at the basal epidermal layer and dermal mucinosis. With the diagnostic hypothesis of an inflammatory myopathy, she was treated with high-dose oral glucocorticoid 1 mg/kg/day, methotrexate (20 mg/week), colchicine (1.0 mg/day), and a calcium channel blocker with a sustained clinical and laboratorial response, except for a minor muscle outbreak at the beginning of the disease. Glucocorticoid was discontinued after three years and

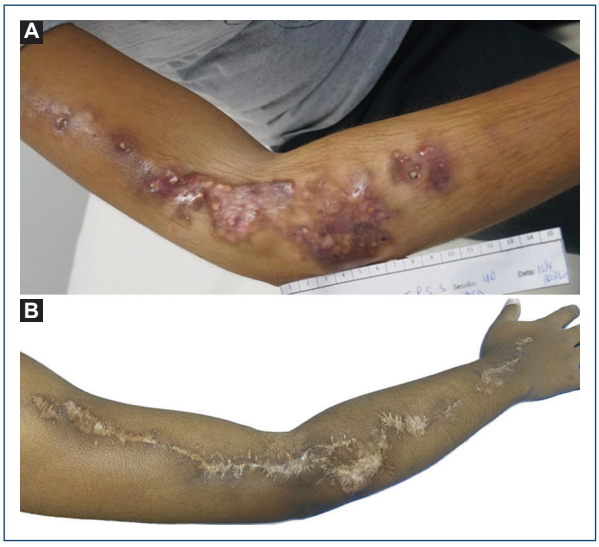

Figure 1. A: cutaneous calcinosis in the right upper limb, where previously was a hypopigmented lesion.
B: appearance of the right upper limb after one year of surgical removal of the calcinosis.

***Correspondence:**
Samuel Katsuyuki-Shinjo
E-mail: samuel.shinjo@usp.br

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methotrexate dose was reduced from 20 to 15 mg/week. There was no further calcinosis and the patient decided to undergo surgical removal of the old ones in 2022 (Fig. 1B). In March 2023 at the age of 20 in our tertiary service, she was clinically and laboratory-stable and specific autoantibodies for myositis were strong positive only for anti-NXP-2 autoantibody (EUROLINE autoimmune myositis 16 Ag, Euroimmun, Lübeck, Germany).

In conclusion, myositis-specific antibodies can contribute to the diagnosis of DM, especially in cases of atypical presentation, as in the reported case.

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Conflicts of interest

None.

Ethical disclosures

Protection of human and animal subjects. The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

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