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CLINICAL IMAGE

Ulcerative rash in a patient with anti-MDA5 antibody-associated clinically amyopathic dermatomyositis

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Presentation

Aman in his sixties presented with recurrent rash for 10 years and joint pain for 3 years. The red macular rash appeared repeatedly on the periorbital, cheeks, ears, chest, neck, and extremities. Recurrent skin ruptures on both auricles and hands persisted for 1 year prior to admission. Physical examination revealed violaceous macules with skin atrophy on the dorsum of his hands, accompanied by skin ulceration (Fig. 1a, b) and red rash with atrophy on both auricles (Fig. 1c, d). Normal muscle strength. Laboratory tests showed anti-MDA5 + , RF + , normal creatine kinase, normal EMG, ANA (-), ENA (-), and ANCA (-). Skin biopsy: small paravalvular lymphocytic infiltrates in the dermis with mucin deposits. CT scan of the chest revealed interstitial changes in both lungs (Fig. 1e). Pulmonary function tests showed mild decrease in diffusion function. He was diagnosed with anti-MDA5 antibody-associated CADM [1]. He was treated with glucocorticoid, cyclosporine, and tofacitinib, and the skin ulcers healed after 2 months.

Discussion

Melanoma differentiation-associated gene 5 (MDA5) antibody is the relatively most common myositis specificity antibody (MSA) in clinically amyopathic dermatomyositis (CADM), the subtype of idiopathic inflammatory myopathy, which is highly correlated with rapidly progressive interstitial lung disease in the Asian population [2]. The disease is always severe, with poor effect of conventional immunosuppression therapies and poor prognoses, which has attracted considerable attention from clinicians in recent years [3]. At present, the pathogenesis of this disease is not clear, and there is no unified standard for the treatment. The rash is the most common symptom of the disease and resembles typical DM, but is often more severe and accompanied by skin ulceration. Multiple hard-to-heal skin ulcers can easily become infected [4]. Cauliflower ear is an unusual presentation of dermatomyositis. This patient's cauliflower ears were due to contraction of the healing scars from repeated skin ulcers. Relapsing polychondritis and ANCA-associated vasculitis were excluded because

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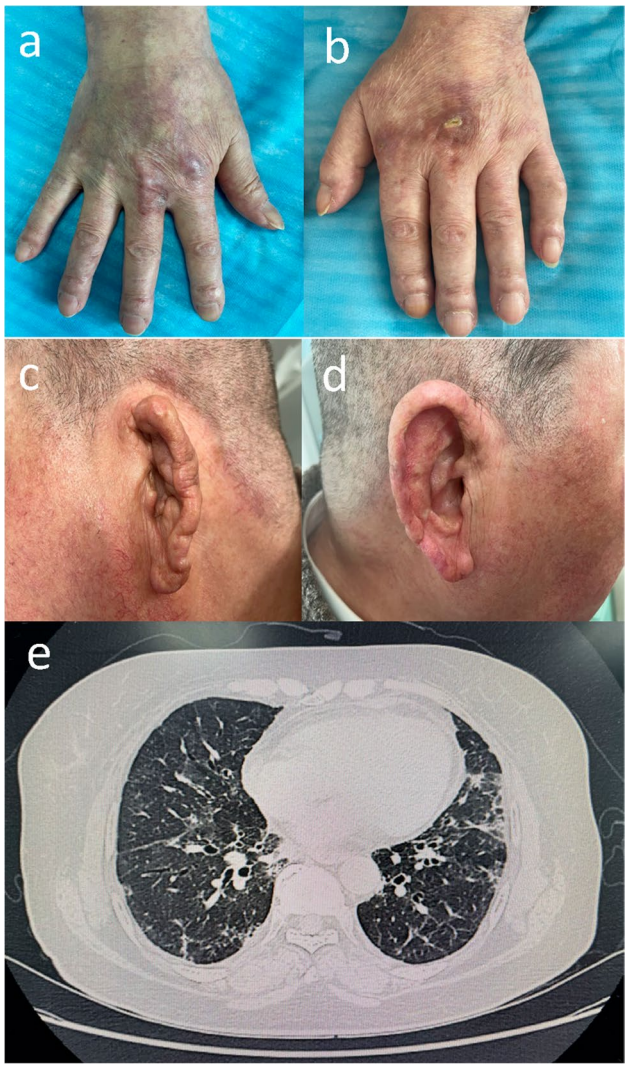

Fig. 1 Anti-MDA5 antibody-associated clinically amyopathic dermatomyositis. (**a**) Violaceous macules with skin atrophy across the dorsum of the right hand. (**b**) Skin ulceration with yellow discharge on the dorsum of the left hand. (**c**) Atrophy with cauliflower-like appearance on the left auricle. (**d**) Red rash with mild atrophy on the right auricle. (**e**) Interstitial lesions in both lungs

there was no organ system involvement such as airway, kidney, and ANCA (-). The patient in this case had skin ulcers as the prominent clinical manifestation, and the interstitial lung lesion was still stable but still at risk of rapid progression. Aggressive treatment was given with good results, but monitoring of the pulmonary lesions is still required [5].

Compliance with ethical standards

Ethics approval This article does not contain any studies with human or animal subjects performed by the author.

Informed consent Verbal consent was obtained from the patient to publish this case and related clinical image. Details that might disclose the identity of the patient were purposely omitted.

Disclosures None.

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