Linear morphea involving the underlying muscle

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Presentation

A 62-year-old woman with hypothyroidism and diabetes visited our hospital with bilateral stiffness and swelling beginning from the forearms and progressing to the fingers. Her symptoms appeared 1 year prior and gradually deteriorated. On physical examination, both forearms showed stiffness and swelling with well-circumscribed glossy skin (Fig. 1a). However, there was no evidence of Raynaud's phenomenon, critical digital ischemia, skin thickening except for upper limbs, dysphagia, dyspnea, or other organ involvement. The results of serum examinations were as follows: creatine kinase (CK), 429 IU/L (normal: 41–153) (CK-MB 27%, CK-MM 73%); aldolase, 7.5 U/L (normal: 2.7–5.9); free-T3, 3.05 pg/mL (normal: 2.20–4.10); free-T4, 0.68 ng/dL (normal: 0.72–1.52); TSH, 7.356 μ IU/mL (normal: 0.400–3.700); random blood glucose, 191 mg/dL (normal: 70–200); and hemoglobinA1c, 6.8% (normal: 4.9–6.0). Antinuclear antibody (ANA) was positive at a titer of 1:80 with homogenous pattern (normal: < 1:40) and anti-single-stranded (ss) DNA immunoglobulin G antibody tested positive at 40.8 AU/mL (normal: < 25). The other antibodies including anti-double-stranded DNA antibody, anti-ribonucleoprotein antibody, anti-topoisomerase I antibody, anti-centromere antibody, anti-RNA polymerase III antibody, anti-aminoacyl tRNA synthetase antibodies, anti-Mi-2 antibody, anti-MDA5 antibody, and anti-TIF-1gamma antibody were negative. Contrast fat-suppressed T1-weighted magnetic resonance imaging (MRI) showed diffuse enhancement of the extensor muscle group, flexor digitorum profundus muscle, and palmaris longus muscle of the right forearm without fasciitis (Fig. 1b). En bloc biopsy of the right forearm revealed increased and thickened collagen bundles in the dermis, and muscle bundles showed

lymphocytic infiltration between the bundles with myonecrosis. The deep fascia did not reveal eosinophilic infiltrates or any inflammation (Fig. 1c, d). She was diagnosed with linear morphea and received combined prednisolone (PSL) 40 mg per day (0.8 mg/kg) and methotrexate (MTX) 8 mg per week, resulting in gradual improvement of stiffness and swelling. Four weeks later, CK and anti-ss DNA were decreased to 57 IU/L and 11.9 AU/mL, respectively, but the weak enhancement of muscles persisted on MRI. Twelve weeks later, PSL was tapered to 10 mg per day, and MTX was increased to 12 mg per week.

Discussion

Morphea, or localized scleroderma, is a rare inflammatory disease characterized by well-circumscribed sclerotic skin changes. Morphea shows a wide spectrum of clinical manifestations, ranging from involvement of the skin and subcutaneous tissue to the musculoskeletal system. Linear morphea presents as band-like cutaneous sclerosis with joint contractures and muscle weakness. Although no specific antibodies are detected in the morphea, ANA is positive in 34%, with a speckled pattern as the most common pattern (81%) on immunofluorescence [1]. However, a homogenous pattern was the predominant (70%) in mestizo Mexican children with morphea [2]. Anti-ss DNA antibody and anti-histone antibody are associated with more extensive skin diseases with joint contractures and reflect disease activity [3]. MRI is recommended for the evaluation of musculoskeletal involvement. En bloc biopsy is useful in detecting the depth of inflammation and differentiating eosinophilic fasciitis, lichen sclerosus et atrophicus, sclerosing panniculitis, and scleromyxedema. Topical corticosteroids and phototherapy are considered for superficial skin lesions, but systemic PSL and MTX treatment should be initiated for deep involvement [4]. We should be familiar with the correlation between the skin and musculoskeletal system.

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Figure 1 **a** Well-circumscribed glossy skin in the left forearm. **b** Diffuse enhancement of the extensor muscle group, flexor digitorum profundus muscle, and palmaris longus muscle of the right forearm on contrast fat-suppressed T1-weighted magnetic resonance imaging. **c** Thickened and increased collagen bundles in the dermis. **d** Lymphocyte infiltration between the muscle bundles with myonecrosis.

Declarations

Consent for publication We obtained written informed consent from the patient for publication.

Disclosures None.

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