Fasciitis Associated with Generalized Morphea

Key words: generalized morphea, MRI, fasciitis, biopsy

Figure 1. Photograph of the legs. A large, thickened, firm, hyper/hypopigmented plaque is evident on the left leg.

Figure 2. Biopsies of affected skin (A) and contiguous tibialis anterior muscle and fascia from the left leg (B–D). (A) The dermal collagen is thickened, packed, and hyalinized. The subcutaneous fat is extensively replaced by collagen, and perivascular inflammatory infiltrates are present (HE stain, ×50). (B) Fibrosis and inflammatory infiltration in the epimysium, the adjacent endomysium, and perivascular sites (HE stain, ×200). (C) Mild variation of fiber size and increased perimysial connective tissue with perimysial and perivascular infiltration (HE stain, ×100). (D) Mononuclear cell infiltration in the superficial layer of the fascia (HE stain, ×200).
A 51-year-old woman reported an eight-month history of multiple cutaneous lesions on the left leg, hip, abdomen, and nuchal region, and wasting of the left leg. A large, thickened, firm, indurated, hypo/hyperpigmented plaque covered the left lower extremity (Fig. 1). Diminished left knee joint mobility and slight ankle extensor weakness were evident. Dermatological evaluation and skin biopsy, revealing sclerotic changes (Fig. 2A), suggested generalized morphea (GM). The left tibialis anterior and extensor digitorum longus muscles were hyperintense on fat-saturated T2-weighted MRI, and gadolinium-enhanced T1-weighted MRI revealed global enhancement of these fascial tissues and thinning of the panniculus anteriorly (Fig. 3). MRI-guided biopsy of the left tibialis anterior muscle and its fascia demonstrated inflammatory infiltration and marked fibrosis in the perimysial and epimysial fascia and in the subfascial region. Adjacent muscle fibers varied in size but necrosis was rare (Fig. 2B–D), while the deep muscle tissue was almost normal. Symptoms improved with oral prednisolone, and MRI normalized after 1 year of therapy.

In GM, a rare subtype of localized idiopathic scleroderma affecting both cutaneous connective tissue and microvasculature, multiple patches of skin sclerosis occur without systemic manifestations (1). In the present patient, MRI clearly detected inflammation along the entire fascial sheet of the muscles underlying the skin lesion, and fasciitis was histologically confirmed. This case demonstrates that widespread deep inflammation can accompany GM, and may need aggressive treatment to prevent joint deformities and skin contracture (2). A careful search for extra-cutaneous lesions is important in therapeutic decision-making and MRI is a useful modality to evaluate and follow deeper inflammation in GM.

References

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