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Case report

Localized scleroderma and regional inflammatory myopathy

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Abstract

Inflammatory myopathy is rare in localized scleroderma. We report 2 new cases of regional inflammatory myopathy associated with localized scleroderma and review 10 reported cases of localized scleroderma associated with an inflammatory myopathy with regional muscle involvement, more often in the upper extremities. Serum creatine kinase was mildly elevated or normal. Histopathology often showed perimysial inflammation and plasma cell infiltration. These cases demonstrate that inflammatory myopathy should be considered in patients with localized scleroderma and regional muscle weakness, pain or atrophy. Muscle biopsy can confirm the diagnosis of myositis, which if identified, will require anti-inflammatory and/or immunosuppressive therapy.
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Keywords: Inflammatory myopathy; Localized scleroderma; Morphea; Linear scleroderma; Perimyositis

Localized scleroderma (LSc) is an idiopathic inflammatory disorder associated with thickening of the skin and subcutaneous tissue in the absence of typical systemic manifestations of systemic sclerosis (SSc), and with better prognosis [1,2]. Depending on the clinical features, LSc is classified as (a) linear scleroderma; (b) circumscribed morphea; (c) generalized morphea; or (d) pansclerotic morphea [2]. Clinical presentation may include sclerotic "band like" skin lesions, which often follow a dermatomal distribution and have hypopigmentation, hyperpigmentation, or subcutaneous and late cutaneous atrophy with a shiny, "paper-like" appearance. In linear scleroderma, the deep subcutis may be affected, and there is rarely underlying bone involvement (melorrheostosis). Extracutaneous manifestations have been reported in up to one-fifth of LSc patients [3], but they differ in type and are not as severe as in SSc. Neurologic complications have been reported in 4% of LSc patients. In the literature, there are only a few well-described cases of inflammatory myopathy associated with LSc (LSc–IM) (Table 1) [4–9].

We describe two cases of LSc with an associated regional inflammatory myopathy, and review the relevant literature. Both of our patients were evaluated by two of us (SAZ and TM). Over the last 3 decades, approximately 500 new childhood and adult-onset patients with LSc have been seen at the University of Pittsburgh scleroderma clinics.

2. Case reports

2.1. Case report 1

A 43-year old Caucasian woman with LSc was diagnosed at 29 years of age when she developed skin

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Table 1
Inflammatory myopathy and localized scleroderma [4–9,20].

Ref	Age[a]/ Gender	Type of LSc	Age at LSc onset (years)	Myopathy after LSc onset (years)	Clinical features	Location of skin changes	CK (IU/L)[b]
[20]	5F	Linear	4	1	Arm weakness, wasting, cramps	Forearm flexor surface, hand	NL
[6]	9 M	Linear	*[c]	7	Arm > leg wasting	Arm	751
[9]	20F	Linear	20	6	Arm, weakness/atrophy/pain	Arm, abdomen	NL
[5]	32F	Morphea	20	12	Arm weakness and atrophy	Forearm, thigh, abdomen	230–457
[8]	33F	Morphea	33	0.4	Bilateral arm weakness	Both arms, chest, back	490
Our1	42F	Linear	29	4	Arm weakness; leg atrophy	Abdomen, shoulder	243
[4]	4 M	Linear/En Coup de Sabre	4	2	Facial hemiatrophy, eye pain, headaches, seizures	n.d.	n.d.
[7]	23F	Linear/En Coup de Sabre	22	2	Facial hemiatrophy, tongue hemiatrophy, dental pain	Face	n.d.
[5]	30F	Linear/En Coup de Sabre	14	2	Hemifacial atrophy, diplopia, bilateral arm weakness	Interscapular, face	NL
Our2	32F	Linear/En Coup de Sabre	11	24	Proximal arm atrophy, weakness; hemifacial atrophy	Shoulder, face	NL

n.d., not described; NL, normal.
[a] Age of onset of myopathy.
[b] In most laboratories serum CK > 200 IU/L is abnormal.
[c] Age of LSc onset is unclear.

thickening in a linear distribution below her right knee, on the right abdomen, and on the right posterolateral thorax. At age 41, she suffered a right tibial fracture followed by right leg atrophy. At age 43, she presented with 8 months of right shoulder weakness that began with the inability to elevate her arm above her head. She denied pain or sensory loss. Examination revealed mild weakness of the right deltoid, triceps, wrist extensors, supraspinatus and quadriceps femoris muscles [all Medical Research Council (MRC) 4+]. There were areas of subcutaneous tissue atrophy and hyperpigmented skin over the right shoulder area near the trapezius muscle and in the right lower abdominal area and right lower extremity, without induration or erythema.

Laboratory testing showed a mildly elevated serum creatine kinase (CK) at 243 IU/L (normal < 200) and a positive antinuclear antibody (ANA) at a titer of 1:640 (normal < 1:40). Serum gammaglobulin was elevated at 2.48 gm/dl (normal 0.70–1.60). Magnetic resonance imaging (MRI) of the lower extremities, cervical and thoracic spine showed asymmetric focal muscle atrophy and fibrosis, suggestive of prior myositis (Fig. 1).

Nerve conduction studies were normal, and needle electromyography showed decreased insertional activity in the right deltoid, rhomboid major and thoracic paraspinal muscles. There were a few voluntary motor unit potentials (MUPs) with decreased durations and amplitudes in the right vastus lateralis muscle and polyphasic configurations in the right deltoid muscle. Needle muscle biopsies were performed on the right deltoid and vastus lateralis muscles. Histopathologic examination of both specimens showed features of a nonspecific inflammatory myopathy (Fig. 2). There were some degenerating and regenerating myofibers, predominantly in the perifascicular regions with random atrophy and hypertrophy. There was perimysial more than endomysial fibrosis, worse in the deltoid. A predominantly perimysial mononuclear cell inflammatory infiltrate included a mixture of CD4- and CD8- reactive T cells, only rare CD123-reactive dendritic cells, macrophages, and plasma cells (CD138+) (Fig. 2A–C). B-cells were not identified via CD20, but there were present via CD79 immunostain. All myofibers showed abnormal, strong immunoreactivity for major histocompatibility complex-1 (MHC-1). Endomysial capillaries did not show membrane attack complex on immunostaining. There was no evidence of capillary loss (via CD31) or vasculitis.

The patient was treated with oral prednisone (up to 20 mg daily), and the dose was gradually tapered off after 18 months. The symptoms improved subjectively, and the serum CK and immunoglobulin levels became normal. There were no clinical or laboratory exacerbations until age 47, when she reported an increase of fatigue, with serum CK elevated at 242 IU/L. She was restarted on prednisone, and methotrexate was added at 15 mg by mouth weekly. The disease remained stable over following 2 years.

2.2. Case report 2

At the age of 11 years, a Caucasian female was diagnosed with LSc “en coup de sabre” after developing right hemifacial atrophy with further progression by age 14. She also had indurated right suprascapular hypopigmented skin areas in a linear distribution. At age