Linear Morphea: A Mimicker of Motor Neuron Disease

Sir,

Even after decades of the initial description of the motor neuron diseases (MND) by Jean-Martin Charcot, understanding the exact pathophysiology and treatment of these disorders is still evolving.[1] Amyotrophic lateral sclerosis (ALS) is the most common form of MND, which is a group of neurodegenerative disorders primarily involving the anterior horn cell along with selective pyramidal degeneration.[2] A diagnosis of MND brings the worst news to patients and their caregivers. Therefore, a clinician must be vigilant to rule out mimickers of MND before confirming the final diagnosis.[3] Herein, we describe a patient in whom linea morphea mimicked the lower motor neuron type of MND.

This 19-year-old male, after being diagnosed with localized MND at another center, visited us for a second opinion. He presented with thinning of the right upper limb and right side of the chest for the past 5 years, which was incidentally noticed by his brother. The thinning initially involved the right forearm and progressed over the subsequent 5 years to involve the arm, right side of the chest, and upper back. Although he was able to perform his daily activities, there was difficulty in lifting heavy objects using the right upper limb. There was no history of muscle twitching, sensory symptoms, or involvement of other three limbs, cranial nerves, or bladder-bowel. Family history was unremarkable. Clinical examination revealed multiple irregular discrete hyperpigmented depressed skin lesions along with diffuse circumferential atrophy over the right forearm, arm, and upper scapular region [Figure 1c-g]. The right-sided face showed subtle atrophy, but there was no thinning or fasciculations seen over the tongue. [Figure 1f]. Whereas muscle power in the right deltoid, biceps, triceps, brachioradialis, wrist flexors, and extensors was Medical Research Council (MRC) grade 4+/5, it was normal in the other limbs. The sensory, cerebellar, and extrapyramidal examination was normal. Whereasright biceps and triceps jerks were 1+, all other deep tendon reflexes were normal.

Laboratory workup including complete blood count, thyroid, liver, and renal function test was normal. Vasculitis workup and gadolinium-enhanced magnetic resonance imaging of the brain and cervical spine were unremarkable. Nerve conduction studies (NCS) revealed reduced amplitude of compound muscle action potential in right axillary and musculocutaneous nerves with preserved latencies and conduction velocities. All other sampled motor and sensory nerves were normal. Electromyogram (EMG) did not reveal abnormal spontaneous activity, and the motor unit action potentials showed normal amplitude, duration, phases, and recruitment. Although our patient had progressive amyotrophy and weakness involving the right upper extremity, the NCS and EMG findings were nonconclusive. Moreover, he lacked any upper motor neuron feature, and his skin lesions suggested a possible secondary etiology, thereby, not fulfilling the Gold Coast criteria for anterior horn cell disease (AHCD).[4] Considering the clinic 0-electrophysiological mismatch, a biopsy from one of the skin lesions was carried out. It showed normal epidermis, thick dermal collagenization with a paucity of adnexal structures along with perivascular inflammation [Figure 1a and b] confirming the diagnosis of morphea.

Due to the lack of any established biomarker for the diagnosis of ALS, the clinician owns the prime responsibility to make

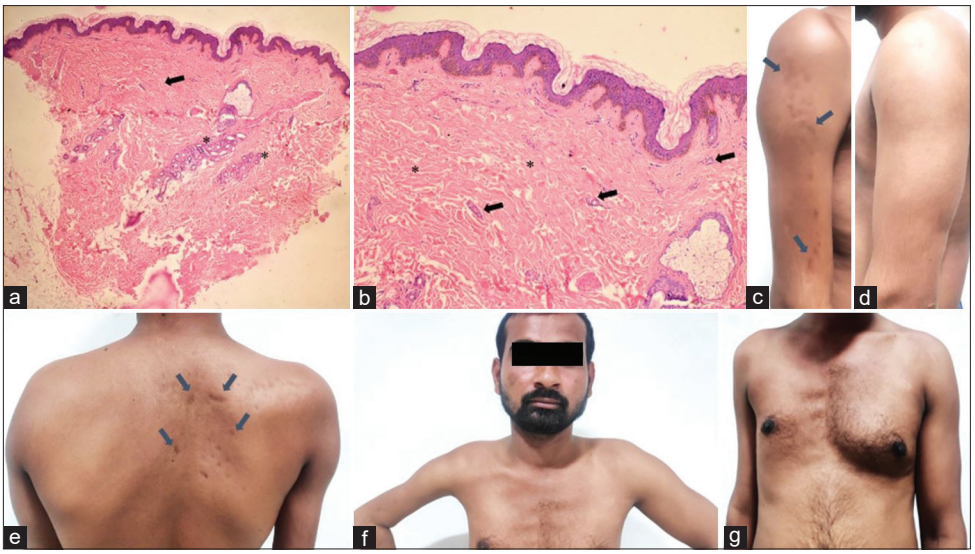

Figure 1: Sections of skin biopsy and photographs of patients. (a) Wedge-shaped skin biopsy exhibiting unremarkable epidermis with dermal collagenization (black arrows) and paucity of adnexal structures (asterisk; H and E, 40 ×). (b) Biopsy shows scant perivascular inflammatory infiltrate (black arrows) and thick collagen bundle (asterisk; H and E, 40 ×). (c-f) Photographs of the patient showing significant wasting of the right upper arm as compared to the left (d and g), along with wasting of the right side of the chest and back with hyperpigmented, depressed scars of burnt-out morphea (blue arrows) over the lateral aspect of the right arm and right upper back (c and e)

the proper diagnosis and differentiate it from mimics. MND can present as classic limb onset ALS, bulbar onset ALS, flail arm variant, flail leg variant, primary lateral sclerosis, or primary muscular atrophy.[2,5] Several disorders like neuralgic amyotrophy, focal chronic inflammatory demyelinating polyneuropathy (CIDP), multifocal motor neuropathy with conduction block, cervical polyradiculopathy, and intramedullary lesions (syringomyelia or tumor) may mimic MNDs.[3] Our patient presented with a primary feature of amyotrophy involving one limb and part of the chest, which may be seen in AHCD, pan-brachial plexus involvement, or pure motor radiculopathy. However, intact sensory system, minimal weakness, and lack of supportive findings in electrophysiology ruled out the involvement of plexus, radicles as well as anterior horn cell. Interestingly, skin lesions were present only over those areas of the limb and chest with associated wasting underneath. Skin biopsy confirmed the diagnosis of morphea in our case.

Morphea or localized scleroderma is primarily an inflammatory disorder affecting the skin as well as underlying subcutaneous tissue leading to sclerosis.[6] A localized form of scleroderma does not cause systemic symptoms like Raynaud's phenomenon, sclerodactyly, pulmonary, renal, gastrointestinal, or cardiac involvement. It can present as plaque morphea, linear morphea, generalized morphea, and mixed morphea.[7] Linear morphea is the most common variant in childhood and adolescence. It can involve the skin, subcutaneous tissue, underlying muscles as well as bone leading to atrophy of the affected part along with deformed growth in children. Only a few cases have been reported with widespread neurogenic atrophy creating a diagnostic dilemma.[8] Other neurological manifestations in morphea include epilepsy, vasculitis, neuropathy, and migraine. Active lesions are defined by the involvement of new areas with violaceous borders and active sclerosis in histopathology. Burnt out lesions are depressed and hyperpigmented as compared to adjoining healthy areas.[7]

Thus, pure motor involvement in the form of progressive mild weakness of a single extremity along with significant amyotrophy of the same extremity as well as the adjoining chest wall in our patient raised a possibility of AHCD. A detailed history along with examination findings of skin lesions and the absence of supportive electrophysiological findings ruled out the possibility of AHCD. If suspected, a biopsy from the skin lesions will confirm the diagnosis of linear morphea.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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