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Clinical Short Communication

Bilaterally symmetric cervical spondylotic amyotrophy: A novel presentation and review of the literature

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ABSTRACT

Background: Cervical spondylotic amyotrophy (CSA) is considered a syndrome of (1) unilateral upper extremity weakness and atrophy, (2) affecting either the proximal or distal musculature, (3) without sensory impairment or lower extremity dysfunction.
Aims of study: The authors report a novel case of bilaterally symmetric CSA with blurring of the proximal–distal distinction, discuss the pathophysiology, and review the literature.
Methods: A 45 year old man presented with a several year history of insidiously progressive bilaterally symmetric upper extremity weakness and wasting, profound in the proximal musculature and moderate to severe in the distal muscle groups.
Results: Based on the clinical, neuroimaging and electrodiagnostic features, this patient harbors a more severe phenotype of the classical syndrome.
Conclusion: The authors propose expanding the generally accepted definition of CSA to include this bilaterally symmetric form of disease, thereby minimizing diagnostic error or delay. Additionally, based on this case and a review of the literature, adherence to the proximal–distal distinction should be avoided since it is commonly blurred. Accurate diagnosis is crucial since this presentation mimics the motor neuron disease variant Vulpian–Bernhardt syndrome. The importance of early diagnosis is underscored by reports of significant improvement with timely surgical decompression.

1. Introduction

Brain et al. first described cervical spondylosis presenting with upper extremity muscle atrophy and weakness in the absence of sensory impairment or myelopathy [1]. Keegan labeled the condition "dissociated motor loss syndrome," Crandall and Batzdorf suggested the term "motor system syndrome," and ensuing reports settled on the phrase "cervical spondylotic amyotrophy" (CSA) [2–4]. This condition is rare, characteristically described as unilateral, and classified according to the most predominantly affected muscle groups as either proximal (scapular, deltoid and biceps) or distal (triceps, forearm and hand) [5]. We present a novel case of bilaterally symmetric proximal CSA with spillover into the distal myotomes, and discuss the pathophysiological implications of this unusual syndrome.

2. Case report

A 45 year old male without significant past medical history or trauma presented to the Tikur Anbessa Hospital Neurology Department in Addis Ababa, Ethiopia with insidiously progressive bilaterally symmetric upper extremity weakness and wasting over the past 6 years. Additionally, he described a two year history of fasciculations in the involved musculature and diffuse intermittent aching of the left upper arm. Family history is negative for neurological disease. He is a government employee, married with seven children, and has no history of alcohol, tobacco or illicit drug use or abuse.

Cognitive functions and cranial nerves were normal. Motor examination demonstrated severe atrophy throughout both shoulder girdles, as well as the bicep and tricep muscles, all being flaccid with spontaneous fasciculations. [Fig. 1] The bilateral forearm and hand muscles demonstrated moderate weakness with atrophy, and there were spontaneous fasciculations throughout the intrinsic hand musculature. Lower extremity motor examination revealed normal tone, bulk and strength. Sensory examination was normal to all primary modalities throughout the upper and lower extremities. Autonomic functions were preserved. Muscle stretch reflexes were

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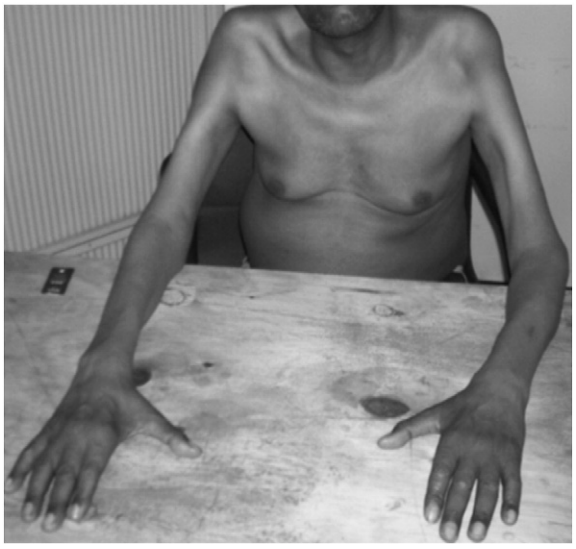

Fig. 1. Severe wasting of the bilateral shoulder girdle musculature and biceps, and moderate wasting of forearms and hands.

hyporeflexic but symmetric in the upper extremities, and normoreflexic in the lower extremities with flexor plantar responses. Stance and posture demonstrated an adducted, hanging position of the arms. Gait including tandem walking was normal except for the absence of armswing.

Routine hematologic studies, CPK and sedimentation rate were normal. HIV testing (ELISA) was negative. Salient features of the nerve conduction studies included mild right ulnar conduction slowing across the olecranon, and bilateral absence of ulnar *F*-waves. Electromyography of the bilateral deltoid, supraspinatus and infraspinatus demonstrated no spontaneous activity and an absence of motor units. Chronic neurogenic motor unit potentials were evident in the bilateral forearms and hand. The trapezius and lower extremity muscles had normal insertional activity, normal motor unit potentials and full recruitment.

Magnetic resonance (MR) imaging of the cervical spine with and without gadolinium demonstrates loss of the normal lordosis with multi-segmental degenerative arthritis and degenerative disc disease, pronounced C3–C7 stenosis, and multi-segmental bilateral foraminal encroachment at C4–C5 and C5–C6, as well as left C3–C4 and right C6–C7 foraminal narrowing. There are multiple levels of marked extradural compression (anteroposterior canal diameters of 10 mm or less at C3–C4, C4–C5, and C5–C6), resulting in significant flattening of the cord with a compression ratio of 0.4. [Fig. 2-A] These findings are pathognomonic for spondylogenic compression of the spinal cord. T2-imaging demonstrates linear, circumscribed non-enhancing high signal intensity (HSI) lesions in the anterior horn (AH) regions consistent with spondylotic myelomalacia. [Fig. 2-B]

3. Discussion

CSA is considered a syndrome with unilateral upper extremity weakness and atrophy of either the proximal or distal musculature, without sensory impairment or lower extremity dysfunction. The proximal variant encompasses a broad phenotypic spectrum including boule musculaire in prodromal or mild stages;[6,7] unilateral wasting of a single muscle;[8] unilateral shoulder girdle wasting, which may occur with distal electromyographic denervation and, in more severe cases, distal weakness;[5,9] unilateral features with electromyographic denervation in the contralateral limb;[10] and unilateral shoulder girdle wasting with mild contralateral weakness [9]. Sporadic case reports have focused on the pathophysiology in order to unravel this clinically heterogeneous disorder [2,8–15].

Keegan, reporting the first autopsy of CSA, demonstrated a normal AH appearance and attributed the pathophysiology to selective intradural ventral nerve root (VNR) compression by posterolateral osteophytes [2]. Matsunaga et al. supported this view by correlating

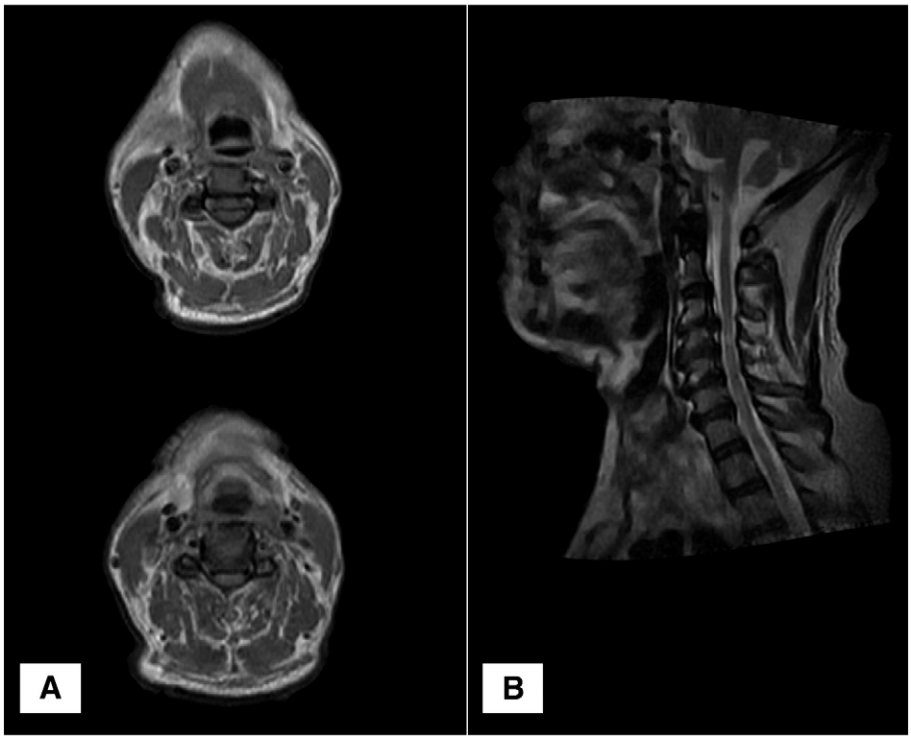

Fig. 2. A. Axial MR images at C3–C4 (top) and C4–C5 (bottom) demonstrating marked extradural compression with a maximal anteroposterior canal diameter of 10 mm, and flattening of the cord with a compression ratio of 0.4. B. T2-weighted sagittal MR image demonstrating severe multi-segmental degenerative spondylotic disease with C3–C7 canal stenosis and circumscribed non-enhancing intramedullary HSI lesions.