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Imaging Case Review

Cervical spondylotic myelopathy in a 68-year-old man diagnosed with amyotrophic lateral sclerosis

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Owing to similar clinical presentations, cervical spondylotic myelopathy can mimic other neurological disorders. In this imaging case review (ICR), we describe a case of cervical spondylotic myelopathy in a patient diagnosed with amyotrophic lateral sclerosis. The key clinical features, imaging findings and differential diagnoses of cervical spondylotic myelopathy compared with amyotrophic lateral sclerosis are also presented.

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KEY WORDS: cervical spondylotic myelopathy; amyotrophic lateral sclerosis; differential diagnosis

Examen du cas par imagerie

Une myélopathie spondylotique cervicale chez un patient de 68 ans atteint de sclérose latérale amyotrophique

En raison de présentations cliniques similaires, la myélopathie spondylotique cervicale peut simuler d'autres troubles neurologiques. Une myélopathie spondylotique cervicale (MSC) chez un patient de 68 ans atteint de sclérose latérale amyotrophique. Les principales caractéristiques cliniques, les résultats d'imagerie et les diagnostics différentiels de myélopathie spondylotique cervicale par rapport à la sclérose latérale amyotrophique sont également présentés.

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MOTS CLÉS : myélopathie spondylotique cervicale; sclérose latérale amyotrophique; diagnostic différentiel

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Case presentation

A 68-year-old man presented to a chiropractic clinic with a two-year history of neck and bilateral shoulder pain, progressive full body weakness, clumsiness of the hands, difficulty with balance and walking, and a previous diagnosis of Parkinson's disease which was refuted by a second neurologist who diagnosed amyotrophic lateral sclerosis (ALS). On physical examination, the patient displayed signs of upper motor neuron lesions (spastic L4 and S1 deep tendon reflexes), lower motor neuron lesions (bilateral C5, bilateral L1-2, and left L4 and S1 motor paresis), and dorsal column and spinocerebellar dysfunction (dysdiadochokinesia in the hands, vibratory sensory loss in the feet, and a wide-based gait), signs consistent with cervical myelopathy. Neck flexion also produced pain and parasthesiae down the patient's spine into his upper extremities (i.e., L'Hermitte's sign). Examination of all 12 cranial nerves, including motor testing of the oculomotor, trochlear, abducens, facial and glossopharyngeal nerves, as well as manual muscle testing of the sternocleidomastoid and upper trapezius muscles, was normal, findings also consistent with a diagnosis of myelopathy.

Magnetic resonance imaging (MRI) of the brain and cervicothoracic spine was taken at a hospital 18 months earlier, after the patient had injured his neck falling backwards off a chair. Brain and brainstem images, including T2-weighted, proton density, and flair-weighted sequences, were unremarkable. Cervicothoracic T2-weighted images revealed multiple levels of compression fractures in the upper thoracic spine in addition to moderate vertebral canal stenosis and effacement of the spinal cord at the C3-4 to C6-7 levels (Figures 1 and 2). These imaging findings together with the lack of clinical bulbar involvement supported the diagnosis of cervical spondylotic myelopathy,[1,2] along with possible primary or secondary spinal neoplasia or other pathologic process (e.g., osteoporosis). The key clinical features, imaging findings, and differential diagnoses for cervical spondylotic myelopathy versus ALS are presented and further discussed in Table 1.

Discussion

According to the revised El Escorial criteria[2,3], the diagnosis of ALS requires evidence of progressive upper and lower motor neuron degeneration compatible with a neurodegenerative disorder that cannot be explained by any other disease process (evident on electrophysiological, imaging, cerebrospinal fluid, or other serological studies)[2]. Investigation results alone, such as evidence of chronic denervation on electromyography, are not adequate for achieving a diagnosis of ALS and must be interpreted with consideration of the patient's history and clinical findings.[2] As such, the patient in our case was referred back to his primary care physician for reassessment and management of cervical spondylotic myelopathy including a recommendation for neurosurgical consultation,[1,4] as well as investigation to rule out primary neoplasia or spinal metastasis. The patient was subsequently referred by his primary care physician for laboratory testing (electrophoresis), but this was negative for plasma cell myeloma.

Four years later (or six years after his initial ALS diagnosis), the patient contracted severe pneumonia and died. In a follow-up telephone conversation between the chiropractor and the patient's wife, it was revealed that the patient was never followed up by his primary care physician for myelopathy and consequently did not undergo spine surgical intervention. He continued to suffer from symptoms of myelopathy including ongoing neck pain and muscle weakness, particularly in the upper extremities, along with bowel and bladder dysfunction (i.e., sensory loss and incontinence), while his bulbar function (i.e., breathing, chewing, swallowing, eye movements, and speech) remained intact, further contradicting a definitive diagnosis of ALS.[1-3,5] Additionally, despite several requests by the chiropractor, copies of the patient's medical and imaging records could not be obtained from the primary care physician. Updated MR imaging of the cervicothoracic spine, if ordered, may have shown progressive deterioration. Other diagnostic methods for differentiating cervical spondylotic myelopathy and ALS are emerging,[6,7] but it is unclear if these were utilized in the current case. For example, levels of cerebrospinal fluid neuron-specific enolase have been shown to be elevated in patients with ALS and as a biomarker can distinguish ALS from cervical spondylotic myelopathy with high sensitivity (0.80) and specificity (0.87).[7] However, it is unknown how the patient in the current case was diagnosed with ALS and whether additional neurological or other conditions (e.g., metastasis) were investigated.

Owing to similar clinical presentations, ALS-mimic syndromes such as cervical spondylotic myelopathy re-

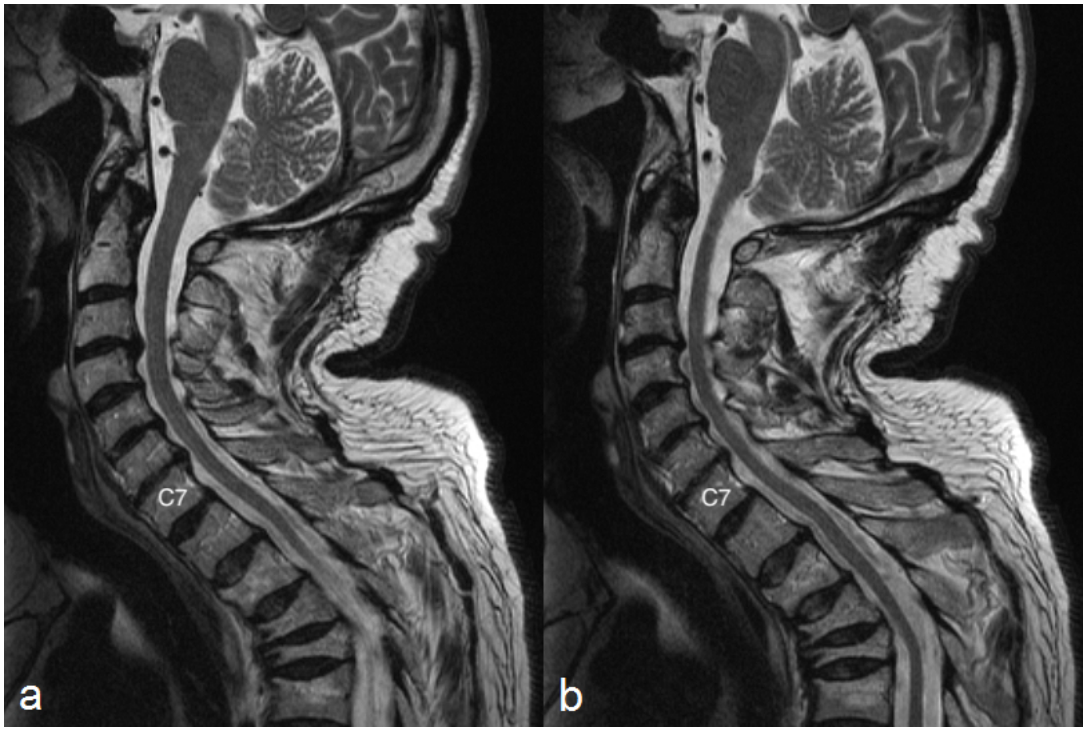

Figure 1.

Right parasagittal (a) and mid-sagittal (b) T2-weighted MR images of the cervical spine without contrast. There is degenerative spondylosis characterized by disc space narrowing and disc contour abnormality. The findings result in moderate vertebral canal stenosis and effacement of the cervical cord at levels C3-4 to C6-7. The Torg-Pavlov ratio ranges between 0.4 to 0.5 at these levels (< 0.8 signifies canal stenosis[9]), further indicating the presence of cervical spinal stenosis. The C7 level is annotated to orient readers to the cervical and thoracic spinal levels. Incidentally noted, there is heterogenous increased signal intensity in the vertebral bodies of T2 to T5 and the T5 superior endplate, indicating normal marrow reconversion. Additionally, there are severe compression fractures consistent with marked compression injuries at levels T3 and T4 and wedge-shaped compression fractures of the T2 and T5 vertebral bodies.

sult in diagnostic error in 5-10% of cases.[2,8] Moreover, the diagnosis of cervical spondylotic myelopathy is often delayed[9], up to an average of 6.3 years in some studies[10], during which time patients' clinical signs and symptoms typically deteriorate[9,10]. As such, clinicians should be aware that cervical spondylotic myelopathy can be confused with, and possibly overlooked in, patients diagnosed with other neurological disorders including ALS. In our case, it remains possible that the patient had diagnoses of both cervical spondylotic myelopathy and ALS. We refer readers to the papers by Wijesekera and Leigh[2] and McCormick *et al*.[9] for additional information on the etiology, diagnosis, clinical management, and long-term prognosis of these conditions.

Key Messages

- Owing to similar clinical presentations, cervical spondylotic myelopathy can mimic ALS in some cases
- Treatable conditions including cervical spondylotic myelopathy should be excluded before ALS is diagnosed

References

1. McCormick WE, Steinmetz MP, Benzel EC. Cervical spondylotic myelopathy: make the difficult diagnosis, then refer for surgery. Cleve Clin J Med. 2003; 70(10): 899-904.
2. Wijesekera LC, Leigh PN. Amyotrophic lateral sclerosis. Orphanet J Rare Dis. 2009; 4: 3.

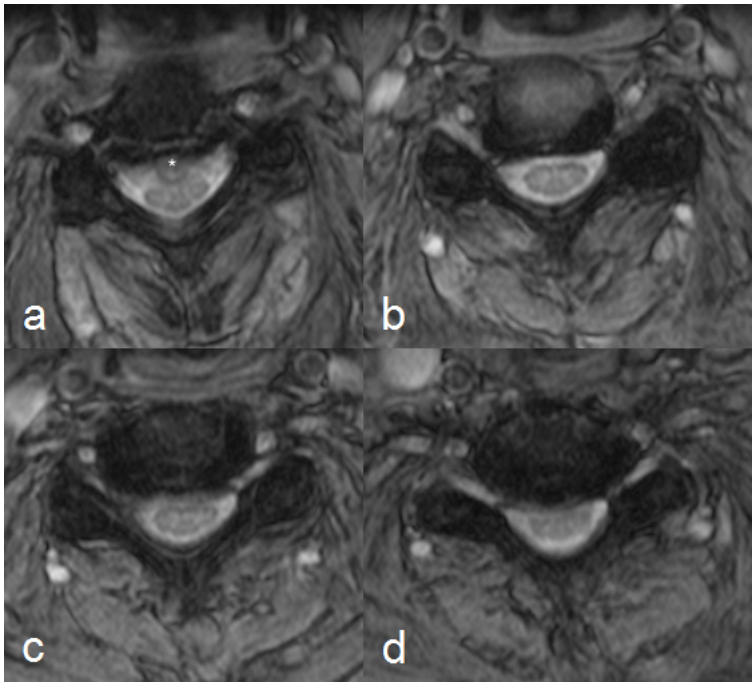

Figure 2.
Axial T2-weighted MR images of the cervical spine without contrast at a) C3-4, b) C4-5, c) C5-6, and d) C6-7. At C3-4, there is a narrow-based posterior central disc extrusion measuring 4.5mm x 3mm (asterisk) causing effacement of the spinal cord. At C4-5 there is disc-osteophyte complex formation with no spinal cord abnormality. At C5-6 and C6-7 there is mild disc contour abnormality with disc-osteophyte complexes causing moderate vertebral canal stenosis and mild effacement of the spinal cord. Other findings include apophyseal hypertrophy and neuroforaminal encroachment demonstrated at all levels.

3. Brooks BR, Miller RG, Swash M, Munsat TL. El Escorial revisited: revised criteria for the diagnosis of amyotrophic lateral sclerosis. Amyotroph Lateral Scler Other Motor Neuron Disord. 2000; 1(5): 293-299.
4. Young WF. Cervical spondylotic myelopathy: a common cause of spinal cord dysfunction in older persons. Am Fam Physician. 2000; 62(5):1064-1070.
5. Kim HJ, Tetreault LA, Massicotte EM, *et al*. Differential diagnosis for cervical spondylotic myelopathy: literature review. Spine (Phila Pa 1976). 2013; 38(22 Suppl 1): S78-S88.
6. Koike Y, Kanazawa M, Terajima K, *et al*. Apparent diffusion coefficients distinguish amyotrophic lateral sclerosis from cervical spondylotic myelopathy. Clin Neurol Neurosurg. 2015; 132: 33-36.
7. Tsukahara A, Hosokawa T, Nishioka D, *et al*. Neuron-specific enolase level is a useful biomarker for distinguishing amyotrophic lateral sclerosis from cervical spondylotic myelopathy. Sci Rep. 2021; 11(1): 22827.
8. Cortés-Vicente E, Pradas J, Marín-Lahoz J, *et al*. Early diagnosis of amyotrophic lateral sclerosis mimic syndromes: pros and cons of current clinical diagnostic criteria. Amyotroph Lateral Scler Frontotemporal Degener. 2017;18(5-6): 333-340.
9. McCormick JR, Sama AJ, Schiller NC, Butler AJ, Donnally CJ 3rd. Cervical spondylotic myelopathy: a guide to diagnosis and management. J Am Board Fam Med. 2020;33(2): 303-313.
10. Sadasivan KK, Reddy RP, Albright JA. The natural history of cervical spondylotic myelopathy. Yale J Biol Med. 1993;66(3): 235-242.

Table 1.
Key clinical features, imaging findings and differential diagnoses of cervical spondylotic myelopathy versus amyotrophic lateral sclerosis.

CERVICAL SPONDYLOTIC MYELOPATHY	AMYOTROPHIC LATERAL SCLEROSIS
Key clinical features[1] • Neck, subscapular, and/or shoulder pain • Upper extremity numbness or parasthesia • Lower extremity sensory (i.e., dorsal column) changes • Upper or lower limb motor weakness • Gait difficulties ("spastic gait") • Upper motor neuron findings (i.e., spasticity, hyperreflexia, clonus, Babinski and Hoffman signs, bowel and bladder dysfunction) • Lower motor neuron findings (e.g., upper limb hyporeflexia and atrophy)	**Key clinical features[3] [a]** • Upper motor neuron signs in the brainstem, cervical, thoracic and/or lumbosacral regions (i.e., clonic deep tendon reflexes [e.g., exaggerated jaw jerk, gag or snout reflexes, Hoffman and/or Babinski responses], pseudo-bulbar features [e.g., dysarthria, dysphagia], forced yawning, spastic facial/upper/lower extremity muscle tone, loss of superficial abdominal reflexes, preserved reflexes in weak wasted limbs) • Lower motor neuron signs in the brainstem, cervical, thoracic and/or lumbosacral regions (i.e., weakness, atrophy, and fasciculations in the jaw, face, palate, tongue, larynx, neck, arm/s, hand/s, diaphragm, back, abdomen, leg/s, foot/feet)
Imaging findings[1,9] • Degenerative spondylosis, including disc space narrowing, disc contour abnormality, posterior disc-osteophyte complex(es), uncinate and articular process hypertrophy • Vertebral canal stenosis and spinal cord effacement • Torg-Pavlov ratio < 0.8 or cervical spinal canal diameter < 12 mm on sagittal imaging • Signal changes in the spinal cord on T2-weighted MR images at the level(s) of spinal cord compression[b]	**Imaging findings[2,3]** • Absence of significant abnormalities of the skull or bones of the spinal canal, brain or spinal cord (suggesting no intra- or extra-parenchymal processes[c], or vascular malformations) on plain x-rays, MR imaging, computed tomography (with or without myelography) or spinal cord angiography that might explain clinical findings • Hyperintensity in corticospinal tracts in the brain, brainstem and/or spinal cord on T2-weighted, proton density-weighted and FLAIR-weighted MR imaging
Differential diagnoses[3,5] • ALS, extrinsic neoplasia (metastatic tumours), hereditary spastic paraplegia, intrinsic neoplasia (tumours of spinal cord parenchyma), multiple sclerosis, normal pressure hydrocephalus, spinal cord infarction, syringomyelia, and vitamin B12 deficiency	**Differential diagnoses[2]** • Cerebral lesions, skull base lesions, cervical spondylotic myelopathy, other cervical myelopathies (e.g., foramen magnum lesions, intrinsic and extrinsic tumours, syringomyelia), conus lesions and lumbosacral radiculopathy, inclusion body myositis, cramp/fasciculation/myokymia syndromes, multifocal motor neuropathy, Kennedy's disease

ALS = amyotrophic lateral sclerosis; *FLAIR* = fluid attenuated inversion recovery; *MR* = magnetic resonance.

[a] A definitive clinical diagnosis of ALS requires the presence of both upper and lower motor neuron signs in the bulbar (i.e., brainstem/cranial motor neuron) region and at least two spinal (i.e., cervical, thoracic, or lumbosacral) regions, or the presence of upper and lower motor neuron signs in three spinal regions.[3]

[b] Signal changes are often but not always present in the cervical cord of patients with cervical spondylotic myelopathy.[1]

[c] Abnormalities confined to the corticospinal tract are consistent with ALS.