We present a case of amyotrophic lateral sclerosis (ALS) in a middle-aged woman who presented with a 2-year history of dysarthria and dysphagia. On examination, she had spastic dysarthria, jaw clonus (video 1), hyperreflexia, and fasciculations involving the tongue, trunk, and extremities. Jaw clonus is an upper motor sign that localizes to the dysfunction of corticobulbar fibers cranial to the 5th nerve nucleus. It is analogous to ankle and patellar clonus in lower limbs. Although jaw clonus is highly suggestive of ALS, it has also rarely been described in patients with multiple cerebral infarcts and CNS demyelinating disorder. Jaw clonus or accentuated jaw jerk serves as a useful clinical sign to distinguish ALS from cervical myelopathy.1,2

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References
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