Cervical spine disease

A possible cause of neuromuscular respiratory failure

Maximiliano A. Hawkes, MD, and Alejandro A. Rabinstein, MD

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Correspondence

Dr. Hawkes
maximilianohawkes@gmail.com

Identifying the cause of neuromuscular respiratory failure is essential for guiding therapy and has major prognostic implications.¹ The most frequent causes can be easily recognized but diagnosing less common etiologies can be challenging.

Case report

A 79-year-old man with chronic cervical spondylosis and radiculopathy requiring 4 decompressive laminectomies and facet injections at C3-4 and C4-5 levels presented with a 9-month history of exertional dyspnea and orthopnea. He described progressive neck pain radiated to both shoulders, worse on the right and accompanied by paresthesias. Recent pulmonary function tests revealed a restrictive respiratory pattern (total lung capacity 76% of predicted, first-second forced expiratory volume 53% of predicted, and forced vital capacity 49% of predicted).

On examination, the patient's neck was stiff and flexed with slight leftward tilt. Passive neck movements revealed limited range of motion and elicited pain radiated to the shoulders. There was no facial or bulbar weakness; tongue appearance was normal. Neck flexors, extensors, trapezius, deltoids, biceps, and triceps were strong bilaterally. Wrist and finger extension were 3/5 bilaterally. There were no fasciculations or fatigable weakness. Both arms were areflexic. Interosseous muscles were hypotrophic bilaterally. The examination was normal in both lower limbs. When lying supine, his abdominal wall moved inward during inspiration (abdominal paradox).

Arterial blood gases on room air revealed a pH of 7.42, PO₂ of 60 mm Hg, PCO₂ of 54 mm Hg, and HCO₃⁻ of 34 mmol/L, consistent with chronic hypoxemic and hypercapnic respiratory failure. Chest film showed elevation of the right diaphragm. Chest CT angiogram and echocardiogram ruled out pulmonary embolism and heart failure. Serum creatine kinase and aldolase were normal.

Sonographic examination of the diaphragm revealed bilateral atrophy, more severe on the right, and paradoxical motion with deep inspiration (figure 1). Nerve conduction studies (NCS) revealed reduced ulnar compound muscle action potentials bilaterally without focal slowing or dispersion. Sensory studies were normal. EMG showed fibrillation potentials in bilateral first dorsal interossei with large motor unit potentials in multiple cervical myotomes, more prominent on the right. Fasciculation potentials were noted in the hand muscles only. NCS and EMG were normal in the legs. Altogether, these findings were consistent with a chronic neurogenic process affecting the cervical myotomes. The relative lack of fibrillation potentials suggested a slowly progressive polyradiculopathy. MRI of the cervical spine demonstrated extensive foraminal narrowing with nerve compression, more

PRACTICAL IMPLICATIONS

Consider diaphragmatic weakness from compressive polyradiculopathy in patients with progressive orthopnea and cervical spondylosis. Nerve conduction studies and EMG, diaphragmatic ultrasound, and MRI are high-yield investigations in this setting.

Department of Neurology, Division of Critical Care Neurology, Mayo Clinic, Rochester, MN.

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Figure 1 Diaphragmatic ultrasound

(A) Expiration, (B) inspiration, (C) expiration. Left hemidiaphragm thickness 0.22 cm at functional residual capacity (A) and 0.16 cm with a paradoxical motion on deep inspiration (B). Right hemidiaphragm thickness 0.11 cm at rest (C) with paradoxical motion. Normal thickness during expiration is 0.22–0.28 cm. End-of-expiration thickness ≤ 0.2 cm indicates atrophy. Thickening $\leq 20\%$ or decreased/paradoxical motion on deep breathing indicates severe weakness.²

prominent on the right, consistent with the predominant right-sided symptoms and greater compromise of the right hemidiaphragm on chest film and ultrasound. The spinal cord was atrophic at C6–C7, and to a lesser degree at C5–6 on the right, with no intramedullary signal abnormality (figure 2).

The patient was started on bilevel positive airway pressure (BiPAP) ventilation with good response, and later discharged home with oxygen supplementation by nasal

cannula during the day and night BiPAP. Given his age and complex surgical history, decompressive surgery was not offered. One year after the diagnosis (22 months after the onset of dyspnea), the patient remains stable. He has no dysarthria, dysphagia, or new weakness in the upper or lower limbs.

Discussion

Cervical spondylosis with compressive polyradiculopathy is an unusual cause of neuromuscular respiratory failure. The phrenic nerve arises from the C3 to C5 spinal nerves. It innervates the diaphragm, which is responsible for 2/3 of the inspiratory effort at rest. While unilateral phrenic nerve palsy is usually compensated by the contralateral hemidiaphragm and the accessory respiratory muscles, cervical spondylosis can cause bilateral phrenic and accessory inspiratory muscles' nerves compression, thus impeding any compensation.

In this case, the patient's history and physical examination were crucial for the correct selection of diagnostic studies. Orthopnea, in the absence of heart failure, and hemidiaphragmatic elevation in the chest film are highly suggestive of diaphragmatic weakness.¹ This was confirmed by the finding of abdominal paradox on physical examination and diaphragmatic ultrasound.² Our patient already had a spirometry before admission; hence bedside pulmonary function tests were not performed. However, they are essential in the initial assessment of neuromuscular respiratory failure. In the appropriate clinical context, a negative inspiratory pressure less than -40 cm H₂O suggests diaphragmatic weakness.¹

Pure respiratory onset and “man-in-a-barrel” motor neuron disease can mimic cervical spine pathology. EMG of the paraspinal muscles can help differentiate the 2 diagnoses. Our patient did not tolerate it, but his persistent asymmetric weakness with sensory symptoms and stable evolution 1 year after presentation make motor neuron disease less likely. Further follow-up will be helpful to rule out this etiology. We cannot entirely exclude that the prior spinal compression, evidenced by localized spine atrophy, could have contributed to the respiratory weakness. However, the absence of respiratory symptoms for more than 20 years after effective surgical decompression and the symptomatic progressive neuroforaminal narrowing, also evidenced by serial MRI, point to cervical radiculopathy as the likely culprit.

Cervical spondylosis with compressive polyradiculopathy is an unusual cause of neuromuscular respiratory failure. It should be suspected in patients with bilateral, asymmetric arm weakness accompanied by sensory symptoms and diaphragmatic weakness. Discrimination from localized forms of motor neuron disease can be difficult and

(A) Sagittal T2-weighted image. Moderate to severe central canal narrowing. Arrow points to spinal cord atrophy. No spinal cord signal abnormality. (B) Axial multiple echo recombined gradient echo-weighted images. Arrows point to bilateral neuroforaminal narrowing at C2-T1, more severe at C3-4. There are no spinal cord signal changes.

eventually hinges on lack of progression or improvement with surgical decompression.

Author contributions

Drs. Hawkes and Rabinstein: study concept, design, data analysis, interpretation, and critical revision. Dr. Hawkes: acquisition of data. Dr. Rabinstein: study supervision.

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Disclosure

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