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A Rare Clinical Presentation - Subacute Respiratory Failure in ALS

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Abstract

Amyotrophic Lateral Sclerosis (ALS) is a progressive motor system disease involving upper motor neurons (corticospinal tract) and lower motor neurons (anterior horn cells). Asymmetrical distal weakness of one limb or bulbar weakness is the typical presentation. Subacute respiratory failure rarely occurs before the diagnosis is established. We present a case of respiratory failure from ALS that appeared after lumbar surgery following a prodrome of "benign" fasciculations.

Introduction

ALS is a motor system degeneration of neurons in the corticospinal and corticobulbar tracts resulting in spasticity and hyperreflexia. The loss of anterior horn cells results in muscle atrophy and weakness. Typically, sensation is spared. Early symptoms depend on the site of first involvement. Common presentations include trip-and-falls from foot drop, clumsy fingers, dysphagia, limb tightness, and muscle cramping [1]. Around-the-clock pattern of disease progression may follow the path of arm, mouth, contralateral arm, and legs. Prognosis varies from months to years. Fasciculations may appear before muscle atrophy and weakness. Respiratory failure is the most common cause of death. We present a case of post-operative respiratory failure that appeared following a prodrome of "benign" fasciculations.

Case presentation

A 76-year-old male presented to our hospital with respiratory failure one month after a lumbar spinal fusion for low back pain. He had been seen at our clinic approximately 8 months prior with fasciculations in his legs and occasional leg cramps. He had unintentional weight loss, without an underlying cause. A diagnosis of benign fasciculations was made when a careful confrontational neuromuscular examination failed to reveal weakness. He had no family history of neurologic disease and no bulbar symptoms or Shortness of breath (SOB). He did not have electrodiagnostic studies prior to his lumbar surgery. He was reassured that without muscular weakness, fasciculations were considered benign. He was asked to have close follow-up.

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Independently, without asking for neurological input, he had lumbar spine surgery at another institution for low back pain. Post-operatively, he developed SOB and hoarseness. He was discharged with home nasal oxygen therapy. He had extensive outpatient evaluation directed by otolaryngology, cardiology, and pulmonary consultants. He had unremarkable imaging of his neck and chest, speech and swallow evaluation, stress test, Holter monitoring and pulmonary function tests.

He presented to our facility with acute worsening of dyspnea. He was dependent on supplemental oxygen since discharge from his lumbar fusion. He developed drooling and inability to swallow solids and liquids. Generalized weakness precluded continued physical therapy. He denied previous similar symptoms, recent changes in medications, paresthesia, or change in bladder or bowel function.

In the emergency department, his breathing was labored requiring 3L nasal cannula to attain an O2 saturation of 94%. His oxygen requirements had increased over the prior 2 weeks with saturations of 70% on the day of arrival. He was dysarthric but oriented and followed commands. Radiographs showed mild bilateral pleural effusions; pCO2 was greater than 104 mmHg (normal 35-45) with serum bicarbonate greater than 45 mEq/L (normal 22-29). He was placed on BIPAP.

A neurologic consultation noted dysarthria with proximal right arm weakness thought secondary to shoulder pain. Muscle bulk, tone, distal power, sensation, and coordination was normal. He had brisk bilateral biceps and triceps reflexes and bilateral non-sustained ankle clonus. Babinski signs were absent. Fasciculations were visible in the bilateral anteromedial thighs.

Twelve hours later, he required intubation. His hospital course was complicated by new onset atrial fibrillation, left lower lobe pneumonia and bronchial mucus plugs.

Acute myasthenia gravis was considered a possible diagnosis so treatment with steroids, pyridostigmine and plasmapheresis was instituted without improvement. A bedside electromyographic test revealed reduced motor unit recruitment in the upper extremities and bilateral proximal and distal denervation (fibrillations and positive sharp waves) in both upper and lower extremities. A diagnosis of motor neuron disease was made.

He declined tracheostomy and was transferred to hospice for terminal extubation with his family at bedside.

Discussion

ALS may be categorized by site of onset: foot drop, thoracic, abdominal, posterior neck, diaphragm, proximal arm/shoulder-girdle, and hemiplegic (*Mills Variant*) [2].

Respiratory failure occurs late in the disease and is the usual cause of death. Only 2.7% of patients presented with early onset respiratory symptoms at a tertiary care outpatient clinic [3].

Just et. al found that simply inquiring about orthopnea in ALS patients could be telling of impending ventilatory failure [4]. Our patient had no preoperative respiratory complaints.

The most recent estimates of ALS prevalence in the United States (2014) were 5 cases per 100,000 [5]. It occurs worldwide with unexplained clustering of cases in Guam and the Japanese Kii peninsula [6].

Caucasians of northern European background are more frequently affected than other racial categories. Onset is in middle age with progression to death in 2-5 years. Men are affected almost twice as often as women [5]. Most cases occur sporadically [7]. Up to 10% are familial with mutations in the superoxide dismutase (SOD1) gene; other rare genetic mutations include TARDBP, FUS, and C9orf72 [2].

The diagnosis can be made based on both clinical and/or electromyographic findings. Electromyography shows re-innervation potentials (i.e. large amplitude polyphasic, long duration motor unit potentials) and denervation (i.e. fibrillations and positive sharp waves) in at least 2 different myotomes. Nerve conduction studies may show involvement of sensory nerves in familial cases [8].

Case reports of post-operative respiratory failure in patients with unsuspected motor neuron disease are few [9,10]. Respiratory insufficiency results from lower motor neuron dysfunction of the diaphragm and accessory muscles of respiration. Symptoms include shortness of breath, orthopnea, sleep disordered breathing, paradoxical breathing, and reduced vocal volume [11]. Preoperative measurement of pulmonary function is unreliable when there is weakness of oropharyngeal musculature.

Our patient had elective lumbar surgery without prior neurological consultation or electrodiagnostic testing. Had widespread changes been detected, he may not have elected to proceed. It is possible the stress of surgery was the trigger that precipitated his rapid decline. Benign fasciculations are “benign” until they are not.

Conclusion

ALS is a progressive neurodegenerative disease which culminates in respiratory failure for most patients. Rarely, respiratory failure may occur before generalized weakness is evident. Benign fasciculations are benign until they are not. If patients with benign fasciculations are contemplating elective surgical procedures, we recommend preoperative screening with electrodiagnostic testing as findings may provide a reason not to proceed.

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