



Managing ALS at the End of Life: An Exploration of Anesthetic Considerations and Palliative Care

Tobenna Mbonu BS¹, Jacqueline Donovan MD¹, Nikita Hansen MD¹

¹Albany Department of Anesthesiology, Albany Medical Center, Albany, NY

Introduction

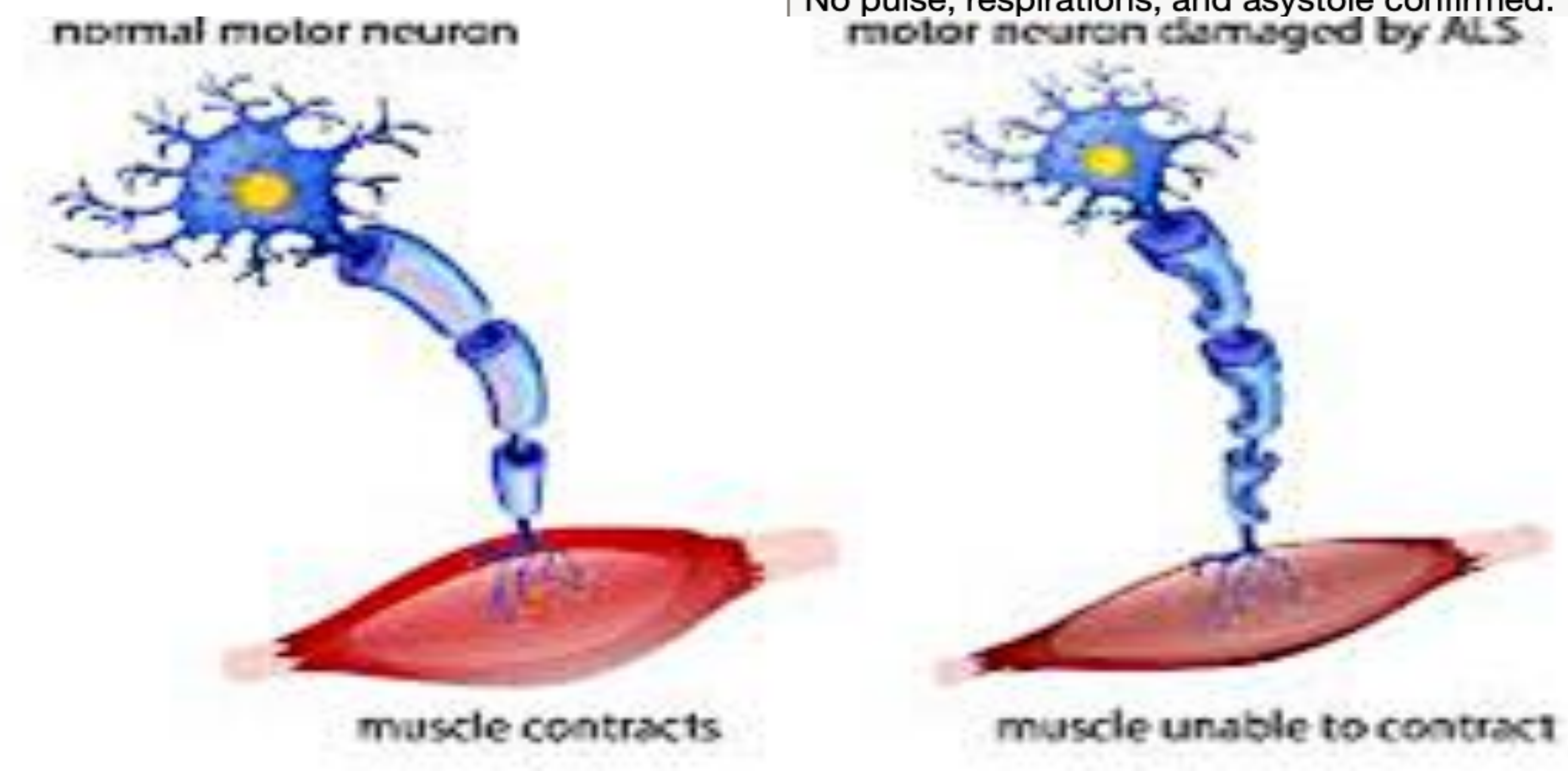
Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease that includes both upper motor neuron and lower motor neuron dysfunction. Lower motor neuron signs can present as flaccid limb weakness, fasciculations, atrophy, and bulbar palsy. Late symptoms can include autonomic symptoms. Life threatening symptoms include respiratory failure and dysphagia that can result in aspiration pneumonia^(1,2). The cause of ALS is poorly understood, but there are familial cases characterized by genetic mutations in genes such as a SOD1⁽²⁾, leading to increased free radical damage to neurons or C9orf72, leading to a loss or gain of function of the resultant protein. Patients with ALS pose unique challenges regarding anesthetic considerations. Here, an eventful case of spinal anesthesia/pain management in advanced ALS can help guide recommendations for future spinal anesthesia use.

Case

A 38-year-old female with an 8-year history of Amyotrophic Lateral Sclerosis (ALS) and DNR/DNI status presented to the emergency department with a 2-day history of RLQ pain, fevers, chills and nausea. CT highlighted recurrent appendicitis, last noted 1 month prior, treated with IV antibiotics. The patient declined surgery at that time for concerns about post-intubation respiratory failure. Medical history included chronic back pain, opiate and benzodiazepine dependence. Physical exam highlighted quadriparesis, reliance on supplemental oxygen, and tachycardia. Pt underwent an open appendectomy, and a combined spinal-epidural (CSE) block was performed as the primary anesthetic in the OR. Under light sedation and continuous monitoring, a spinal dose of 3ml of 0.5% bupivacaine at L3-L4 was placed. Patient tolerated the procedure with no immediate complications. During the anesthesia post evaluation, airway patency and spontaneous ventilation were determined. A multimodal analgesia pain management approach was initiated.

Hospital Course

Stage	Details
Presentation	Patient with ALS presented with appendicitis. Prior appendicitis treated with IV antibiotics; opted out of surgery due to ventilation dependency risk with intubation.
Surgical Decision	Appendectomy recommended due to recurrence and antibiotic failure. Surgical approach changed from laparoscopic to open after discussion of patient's goals of care emphasized avoiding general anesthesia/ intubation at all costs
Open Appendectomy	Procedure completed under spinal anesthesia; patient moved to progressive care unit.
Post-Operative Complications	blood in JP drain, worsening RLQ pain, persistent tachycardia
CTA and Embolization	Imaging showed hemoperitoneum in RLQ with active bleeding from appendiceal artery. Embolization performed; transferred to SICU.
Progressive Respiratory Failure/Pain Management	Required BiPAP; Pain managed with Dilaudid (0.5 mg/hr) and Ketamine (0.05 mg/kg/hr).
Worsening Shock	Condition worsened despite vasopressors. Goals of care discussed with family
End of Life	Heart rate and blood pressure declined to cardiac arrest. No pulse, respirations, and asystole confirmed.



Discussion

This case sheds light on the complexities of anesthesia and palliative care in patients with advanced ALS, especially those with DNR/DNI status. The use of neuraxial anesthesia was innovative and critical in avoiding risks associated with general anesthesia, such as worsened respiratory function/ residual muscle paralysis requiring ventilation support. The case elucidates the importance of an individualized approach to anesthetic planning and the incorporation of palliative care in managing complex cases. This balance between acute surgical needs and advanced directives spotlights the need for comprehensive preoperative discussions.

Conclusion

A patient will ALS safely underwent neuraxial anesthesia with mild sedation, maintaining spontaneous ventilation and avoiding airway manipulation, making spinal anesthesia a feasible alternative to general. Pain management in ALS patients, who can experience chronic opioid dependence, poses more challenges. A multimodal analgesia approach was used, including a ketamine infusion. to address the extreme pain.

References

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