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Case Report

The Safe and Effective Use of Lumbar Epidural for the Surgical Management of a Femoral Neck Fracture in a Patient with Progressed Amyotrophic Lateral Sclerosis (ALS): A Case Report

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Abstract

Objective: Because of the limited publications on the appropriate anesthetic management of patients with Amyotrophic Lateral Sclerosis (ALS), this manuscript is a means of providing outcome data on the use of epidural anesthesia in an ALS patient and to discuss the risks and benefits of neuraxial anesthesia in these individuals. ***Background:*** ALS is a debilitating neuromuscular disease that arises from destruction of large upper and lower motor neurons. Patients develop weakness of their peripheral muscles that can progress to include intrinsic muscles of the airway and oropharynx, which creates a challenge when determining the safest anesthetic plan and considerations for the post-operative course. ***Case:*** The case discusses a 78-year-old female with a longstanding history of ALS, for which she required noninvasive mechanical ventilatory support and gastrostomy tube feedings secondary to oropharyngeal and upper respiratory muscle weakness and atrophy. The patient presented for orthopedic repair of a femoral fracture. She underwent epidural anesthesia successfully without any exacerbation of her lower extremity weakness. ***Conclusion:*** Regional anesthesia in ALS patients appears to be a safe means of delivering intraoperative anesthesia and postoperative analgesia without compromising baseline functional status. This may be a superior technique to general anesthesia and endotracheal intubation, particularly in patients who already suffer from dysphagia and respiratory compromise.

Keywords: ***lumbar, epidural, neck fracture, als, anesthestia***

Introduction

Disease Process

Amyotrophic Lateral Sclerosis (ALS) is a fatal progressive neurodegenerative disease that affects the upper and lower motor neurons of the brainstem, corticospinal tract, and anterior horn of the spinal cord, respectively.[1,2] Patients affected by this disease usually present with gradually worsening focal weakness and muscle atrophy, typically beginning in the distal extremities such as the hands. It then progresses to proximal muscle groups that affect speech and truncal muscles, usually leading to dysarthria, atrophy of the tongue, as well as respiratory failure from weakening of the diaphragm and accessory muscles.[2] These patients may also suffer from spasticity and hyperreflexia as is expected with upper motor neuron involvement.[1]

The incidence of ALS is 2 in every 100,000 and it is sporadic in nature.[1] There is currently no available curative treatment for this disease. Patients affected by this devastating illness usually die within 3-5 years of diagnosis.[3,4] Extensive research and clinical trials for understanding this disease process and potential therapies have been implemented as our population's life expectancy continues to increase. Although the exact mechanism of the disease is not thoroughly understood, there has been an association with a mutation in the superoxide dismutase protein.[1] Other etiologies implicated in the disease process involves the destruction of motor neurons from excessive levels of the glutamate.[5] The excitotoxicity of the glutamatergic neurotransmitter system has led to the development of a glutamate-release inhibitor, Riluzole, the only disease-modifying agent currently available and FDA approved to treat ALS.[1,5] This neuroprotective agent has been shown to prolong survival for about three to four months, but no definitive treatment is available, and management is usually supportive in nature.[4]

Anesthetic Considerations

Because of the nature of this disease process, primary research of perioperative management in ALS patients is minimal. A

randomized controlled trials would raise both ethical and logistic concerns with barriers in obtaining an adequate sample size. Thus, the majority of our anesthetic planning is secondary to case reports or general knowledge of the properties of the anesthetics administered.[1]

Use of neuromuscular blocking drugs (NMBDs) in patients with any musculoskeletal disease is of concern. In particular, the use of succinylcholine in ALS can result in hyperkalemia from denervation and chronic immobility. While the use of nondepolarizing neuromuscular blockers can result in prolonged immobility and exacerbate weakness, making anesthesia providers cautious of neuromuscular blockade in these patients. Furthermore, utilizing agents that can depress respiratory drive in individuals with an already compromised respiratory status, such as sedatives and analgesics, can pose a great threat in the perioperative phase. This includes increased risk of apnea or aspiration, particularly in patients with pre-operative bulbar involvement.[2] However, axial and regional techniques also carry a potential of worsening patient's functional status if nerve injury were to occur.

It is imperative to determine the safest and most effective management. This depends on a comprehensive approach with the development of an appropriate anesthetic plan for the required surgical procedure, while attempting to mitigate the potential complications associated with our interventions.

Patient Case

78 year old female with a past medical history of ALS presented to our emergency department with a left femur fracture after sustaining a fall at home while ambulating with her walker. The orthopedic department evaluated the patient and determined that an intramedullary nailing of the femoral fracture would be the appropriate treatment.

Her preanesthetic evaluation revealed a rather frail female in the advanced stage of her ALS, who at a height of 162 centimeters, weighed only 38 kilograms. She was dysarthric and her dysphagia had required her transition to enteral feeds through a gastrostomy tube as oral intake was no longer a safe means of nutrition. Despite being able to breathe independently during the day while sitting and standing, she was too weak to inspire adequately when supine, and was thus reliant on noninvasive mechanical ventilation while asleep. This allowed her adequate tidal volumes of about 700-1300 mL, at a rate of 12 breaths per minute with a cough assist mechanism to minimize hypercapnic narcosis from diminished respiratory effort.

Because of her severe bulbar involvement and the potential of altogether losing her already diminished airway tone, we attempted to avoid any invasive airway manipulation that would further exacerbate her respiratory weakness and prolong post-operative mechanical ventilation. Keeping her spontaneously ventilating with an intact respiratory drive was of great priority in our anesthetic plan. We thus turned to a neuraxial approach with an epidural and we felt that this would be a safer approach given her functional status. Because she was ambulatory with assistance at baseline, we felt her peripheral motor function was relatively preserved. It was decided that performing epidural anesthesia would confer the greatest safety profile, while also allowing for adequate post-operative pain control. After much discussion with the patient about the different anesthetic types and the various risks each carry, we proceeded with a regional anesthetic.

After appropriate consent was obtained, the patient was taken to the operating room. All ASA monitors were applied per protocol and the patient was placed in the right lateral decubitus position. Her lumbar interspinous space at the L3-L4 level was located and demarcated. Under the appropriate sterile techniques and with adequate infiltration of local anesthetic into the skin, subcutaneous tissue, muscular, and prespinous space, a 17 gauge Touhy needle was used to access the epidural space at approximately four centimeters via a loss of resistance to air technique. The catheter was advanced slowly and without any resistance to about 9 centimeters at the skin. A test dose consisting of 3 mL of 1.5% Lidocaine with 1:200,000 epinephrine was infused and negative for intravascular placement. After appropriate and sterile securement of the catheter, the patient was placed in the supine position with slight elevation at the head of the bed for improved respiratory effort, and her oxygenation was maintained with 6L of oxygen via a facemask. Seven milliliters of 2% Lidocaine was infused into the epidural and sensory testing after seven minutes indicated she had obtained an adequate anesthetic level to about the T10 dermatomal level.

For adequate amnesia, we elected to administered small incremental doses of Ketamine to preserve a patent spontaneously ventilating airway. The patient tolerated the procedure remarkably well; she maintained stable vital signs, adequate analgesia, amnesia, and akinesis throughout the course of the surgery. Upon completion, she was taken to PACU where a full neurological examination was performed as the epidural local anesthetic effect cleared. After confirming return of her motor and sensory function to her baseline, the epidural catheter was utilized for post-operative analgesia, minimizing the amount of systemic opioids that could cause apnea. She remained comfortable with an infusion of Bupivacaine 0.0625% at 10 milliliters per hour, which was kept for approximately three days until her discharge to a rehabilitation center.

Discussion

The anesthetic management for patients with ALS remains controversial due to limitations in conducting primary prospective studies. However, despite the limited available publications, there have been several reported accounts regarding management of patients with this disease. As expected, case reports have suggested that the use of neuromuscular blocking agents during general anesthesia in patients with ALS results in prolonged motor blockade.[6] Although there is an adequate and appropriate response to the anticholinesterase reversal agents and newer drugs like Suggamadex, the risk of residual weakness and inability to extubate post-operatively increases pulmonary complications.[6,7] Minimal use of NMBDs even in general endotracheal anesthesia can be accomplished with larger doses of inhaled and intravenous anesthetics, and opioids during both airway manipulation and for maintenance of muscle relaxation. This appears to be a successful and potentially safer alternative for neuromuscular blockade.[8,9]

However, general anesthesia is not mandatory to achieve analgesia and akinesis, and can be accomplished with a neuraxial approach in such patients.[10] Several publications have suggested the safe use of epidural anesthesia for the management of ALS patients, even in those undergoing abdominal surgeries while maintaining spontaneous ventilation and sedation.[2,10]

Although there is no one appropriate means to administer anesthetics to ALS patients, regional and neuraxial techniques appear to be favorable when feasible. Pursuing a minimally invasive approach to such patients in order to sustain spontaneous ventilation helps avoid the risk of post-operative ventilator support and the numerous complications that arise with prolonged intubation, such as pneumonia and weakening of the diaphragm.

This is particularly true for patients who already have compromised airway protection secondary to bulbar involvement. We recommend and encourage that regional anesthesia be instituted comfortably in the ALS surgical patient, if possible, particularly in patients with advanced disease.

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