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Anesthetic Management in a Parturient with Advanced Amyotrophic Lateral Sclerosis Undergoing Dilation and Curettage

*Benjamin P Morey, MD, Hong Liu, MD and Cristina Chandler, MD**

Department of Anesthesiology and Pain Medicine, University of California Davis Health, Sacramento, California, USA

Abstract

Background: Amyotrophic lateral sclerosis (ALS) is a rare but debilitating neuromuscular disorder that is characterized by degeneration of both upper and lower motor neurons and successive muscle denervation. More than 50% of patients pass away within 3 years of symptom onset from fatal respiratory muscle failure. Respiratory decompensation in a pregnant patient with ALS is of particular concern given the substantial physiologic increase in minute ventilation that starts early in pregnancy.

Case presentation: 38-year-old female with medical history of advanced ALS, asthma, prior substance abuse, and recurrent urinary tract infections presented with right-sided abdominal and flank pain and 13 weeks of incident pregnancy. The patient was quadriparetic at baseline, had significant bulbar symptoms, and required BiPAP at home. After discussion with a multidisciplinary team of experts, the patient decided to terminate the pregnancy given the substantial risk of exacerbating her already debilitating ALS symptoms. The patient received a combination of para-cervical local block and monitored anesthesia care (MAC). The procedure was without incident, and the patient was discharged home the next day after an uneventful post-operative course.

Conclusion: It is strongly recommended maintaining a steadfast commitment to patient safety and avoiding general anesthesia in ALS patients unless there are irrefutable contraindications to MAC, regional block or neuraxial anesthesia.

Keywords

Amyotrophic lateral sclerosis, Neuromuscular blocking drugs, Monitored anesthesia Care

Background

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease that affects the lower motor neurons originating from the anterior horn of the spinal cord, in addition to the upper motor neurons that comprise the corticospinal tracts. Patients classically display signs and symptoms consistent with disease of both upper motor neurons (muscle spasticity/ stiffness, hyperreflexia) and lower motor neurons (atrophy, fasciculations) [1,2]. The management of patients with ALS presenting for urgent or emergency procedures in addition to elective operations such as tracheostomy or gastrostomy tube placement represents a challenging task for the anesthesiologist. General anesthesia is problematic given the possibility of pulmonary aspiration, exaggerated responses to non-depolarizing neuromuscular blocking drugs (NMBDs), risk of hyperkalemic cardiac arrest with succinylcholine, and concern for postoperative respiratory depression [2]. Moreover, there has been concern for decades that neuraxial anesthesia could potentially exacerbate neuromuscular disorders, which has subsequently led to hesitation when considering spinal or epidural anesthesia in these patients [3]. Here, we describe the anesthetic management of a patient with advanced ALS undergoing a dilation and curettage (D&C) for pregnancy termination, and further discuss the anesthetic implications and physiologic challenges for the parturient with ALS. Written informed consent was provided by the patient for the publication of this case.

Case Presentation

Patient is a 38-year-old G6P4014 woman who presented to the emergency department with right-sided abdominal and flank pain. Her medical history was significant for advanced ALS, asthma, prior substance abuse, and recurrent urinary tract infections. She was diagnosed with ALS 8 years prior. Her symptoms began with paresthesias in the right hand that progressively spread to the rest of her limbs. Within several months, the patient began to experience disequilibrium and weakness in the lower extremities resulting in several falls. She was eventually diagnosed with a slowly-progressive form of ALS, and at the time of presentation was quadriparetic and reliant on a full-time caregiver. The patient also had debilitating bulbar symptoms (dysphagia and dysarthria) and restrictive lung disease requiring intermittent bilevel positive airway pressure. Her regular medication included clonazepam, ropinirole, amitriptyline, and albuterol. After evaluation, the patient was diagnosed with pyelonephritis and subsequently

admitted to hospital. An ultrasound performed at the time of admission revealed the patient to be 13 weeks pregnant. After a lengthy discussion with the patient on the inherent risks of pregnancy given her advanced ALS, she made the decision to terminate the pregnancy.

Preoperative examination demonstrated a weak cough with decreased breath sounds in both lung bases. Patient's blood pressure was 90/40 mmHg with sinus tachycardia 110-112/min. Blood serum tests were normal except for a high white blood cell count 15.0 × 109/L and low hemoglobin 10 grams /dl. She was febrile at 38 degrees Celsius, and described a flank pain with a pain score of 5/10.

The patient had no prior anesthetic history since her diagnosis of ALS. She categorically refused any central neuraxial procedure, and we decided against general anesthesia and endotracheal intubation with neuromuscular agents for its evident risk. LMA was excluded because of bulbar dysfunction. Therefore we used a combination of a paracervical block and MAC. She reported no dyspnea and maintained an oxygen saturation > 95% while being provided supplementary oxygen in supine position. The patient's ventilation was monitored via end-tidal carbon dioxide. Intraoperatively, she received 1 mg midazolam and 25 mcg of fentanyl via a 20 gage peripheral venous line prior to the placement of a paracervical block using 30 mL of 1% lidocaine injected at ~ the "3 and 9 o'clock" positions of the cervix. She also received a total of 24 µg of dexmedetomidine in intermittent 4 µg doses for sedation. The procedure was uneventful and patient was very satisfied. She received 1g acetaminophen in post-anesthesia care unit (PACU) and was discharged home the next day.

Discussion

As demonstrated in this patient, ALS typically begins with focal weakness in the extremities, with subsequent involvement of the proximal musculature. The time to diagnosis as measured from symptom onset is ~ 12 months [1]. While muscle weakness in the extremities is perhaps the most recognizable manifestation of ALS, the most debilitating is likely the bulbar symptoms about 1/3 of patient experience [1]. These include dysarthria, dysphagia, and recurrent pulmonary aspiration that can lead to significant morbidity. Behavioral changes are also commonplace and are the result of a pseudobulbar palsy characterized by considerable emotional lability including episodes of both laughter and crying in response to nominal stimuli. The ALS usually has a rapidly progressive course, with a > 50% mortality rate within 3-5 years of diagnosis as a result of respiratory complications [1,2].

About 10% of ALS cases are familial in nature and generally displaying an autosomal dominant pattern of inheritance [1,2]. The remainder of cases are the result of sporadic mutations. At present, riluzole and edaravone remain the only therapies for ALS approved by the United States Food and Drug Administration (FDA) have demonstrated an extremely limited survival benefit [1]. Thus, the management of these patients continues to rely heavily on symptomatic improvement and the prevention of adverse events. Enteric feeding, aspiration prevention, and ventilatory support (usually bilevel positive airway pressure) are routinely employed.

Descriptions of parturients with ALS are scarce, not least because the disease most frequently affects males in the 5th to 7th decades of life [1]. Nonetheless, pregnancy represents a potentially dangerous physiological challenge to the ALS patient. This is primarily due to the increased ventilatory requirements needed throughout pregnancy due to significant neuromuscular weakness. Most women with ALS who go on to deliver at term have reported no apparent influence on fetal development or infantile morbidity [4-7]. However, while many case reports document an unaltered disease course in parturients with ALS, there are several documenting a precipitous decline in respiratory function and increased mortality. Chiò and colleagues documented one such case in which a patient diagnosed with ALS during the 3rd month of her pregnancy experienced rapid progression of weakness ultimately resulting in interruption of the pregnancy and death from respiratory failure [4]. This has led many experts to advise patients with advanced ALS to forego childbirth altogether.

While the successful application of general and neuraxial anesthesia has been described, there remains a degree of risk with both. Postoperative respiratory depression is the major concern surrounding general anesthesia, with worsening neurologic injury being a theoretical concern associated with neuraxial techniques. The risk of prolonged neuromuscular weakness when employing non-depolarizing NMBDs is especially concerning. However, this has been negated both via complete avoidance, as well as through the use of sugammadex for reversal of neuromuscular blockade [8,9]. In spite of this, patients with ALS still remain vulnerable to the respiratory depression induced by other agents commonly used in a balanced anesthetic plan.

Neuraxial techniques have been avoided for decades in patients with ALS given the concern for potential worsening of neurologic outcomes. This recommendation was first documented by Dripps and Vandam in 1956 who reported exacerbation of several different neurologic disorders in patients who had received neuraxial anesthesia [3]. Potential explanations for this observation include direct injury from the needle or catheter, local anesthetic toxicity, and ischemia from

adjuncts combined with local anesthetics [10]. However, more recent evidence has suggested that those early reports are less concerning than previously thought. In accordance, there are multitude of case reports documenting the successful employment of neuraxial anesthesia in ALS patients without any apparent adverse effects or disease progression [11-13].

To the best of our knowledge, there are no descriptions detailing the use of MAC and paracervical local anesthesia in a patient with ALS undergoing D&C. As a neuraxial technique was firmly declined by the patient, our anesthetic plan sought to avoid the adverse respiratory effects associated with general anesthesia. The reliance on dexmedetomidine as the primary sedative was critical given its lack of respiratory depression [14]. We felt it superior to ketamine (another drug with a favorable respiratory profile) given ketamine's association with salivary secretions (especially concerning in patients at risk of aspiration) and emergence reactions. MAC with propofol and higher doses of opioids was not preferred due to the risk of deep sedation in the setting of major risk of aspiration in a patient with bulbar symptoms.

In conclusion, despite multiple documented positive outcomes with general and neuraxial anesthesia in ALS patients, neither of them is risk-free. While MAC remains the most common anesthetic approach to a D&C, general or neuraxial anesthesia is still utilized on a case-by-case basis. Although far from robust evidence given the small number of patients, there have been few complications when performing neuraxial techniques in patients with ALS. Thus, we feel it is reasonable to offer spinal or epidural anesthesia to ALS patients provided they are aware of the theoretical, but ultimately unknown and unquantifiable risk of neurologic injury. However, we strongly recommend maintaining a steadfast commitment to patient safety and avoiding general anesthesia in ALS patients unless there are irrefutable contraindications to MAC, regional block or neuraxial anesthesia.

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References

1. Brown RH, Phil D, Al-Chalabi A. Amyotrophic Lateral Sclerosis. N Engl J Med. 2017; 377: 162-72.
2. Prabhakar A, Owen CP, Kaye AD. Anesthetic management of patient with amyotrophic lateral sclerosis. J Anesth. 2013; 27: 909-18.
3. Dripps RD, Vandam LD. Exacerbation of pre-existing neurologic disease after spinal anesthesia. N Engl J Med. 1956; 255: 843-9.
4. Chiò, A., Calvo, A., Di Vito, N., Vercellino, M., Ghiglione, P., Terreni, A., Mutani, R., & Mora, G. Amyotrophic lateral sclerosis associated with pregnancy: report of four new cases and review of the literature. Amyotroph Lateral Scler Other Motor Neuron Disord. 2003; 4: 45-8.
5. Leveck DE, Davies GA. Rapid progression of amyotrophic lateral sclerosis presenting during pregnancy: a case report. J Obstet Gynaecol Can. 2005; 27: 360-2.
6. Huston JW, Lingenfelder J, Mulder DW, Kurland LT. Pregnancy complicated by amyotrophic lateral sclerosis. Am J Obstet Gynecol.1956; 72: 93-9.
7. Lupo VR, Rusterholz JH, Reichert JA, Hanson AS. Amyotrophic lateral sclerosis in pregnancy. Obstet Gynecol. 1993; 82: 682-5.
8. Xiao W, Zhao L, Wang F, Sun H, Wang T, Zhao G. Total intravenous anesthesia without muscle relaxant in a parturient with amyotrophic lateral sclerosis undergoing cesarean section: A case report. J Clin Anesth. 2017; 36: 107-9.
9. Yoo JH, Kim SI, Park SY, Jun MR, Kim YE, Kim HJ. Use of sugammadex in a patient with progressive muscular atrophy and in a patient with amyotrophic lateral sclerosis. Medicine (Baltimore). 2017; 96: e7098.
10. Kopp SL, Jacob AK, Hebl JR. Regional Anesthesia in Patients With Preexisting Neurologic Disease. Reg Anesth Pain Med. 2015; 40: 467-78.
11. Hara K, Sakura S, Saito Y, Maeda M, Kosaka Y. Epidural Anesthesia and Pulmonary Function in a Patient with Amyotrophic Lateral Sclerosis. Anesth Analg. 1996; 83: 878-9.
12. Hobaika AB, Neves BS. Combined spinal-epidural block in a patient with amyotrophic lateral sclerosis: case report. Rev Bras Anestesiol. 2009; 59: 206-9.
13. Kusakai M, Sawada A, Kii N, Tokinaga Y, Hirata N, Yamakage, M. Epidural anesthesia combined with sedation with dexmedetomidine for appendectomy in a patient with amyotrophic lateral sclerosis: a case report. JA Clinical Reports. 2018; 4: 82
14. Bohringer C and Liu H. Is it time for an expanded role of dexmedetomidine in contemporary anesthesia practice? – A clinician's perspective. Transl Perioper & Pain Med. 2018; 5: 55-63.

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***Corresponding Authors:** *Cristina Chandler, MD, Associate Professor, Department of Anesthesiology and Pain Medicine, University of California Davis Health, 4150 V Street, Suite 1200, Sacramento, CA 95817, USA, Tel: 916-734-5031, Fax: 916-734-7980, E-mail: cmchandler@ucdavis.edu*

Editor: *Li-ming Zhang, MD, Associate Professor, 200 Lothrop Street, UPMC Montefiore, Anesthesiology, Suite 467.2, Pittsburgh, PA 15213, USA, E-mail: zhangl1@anes.upmc.edu*

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