Anesthetic management of a parturient with amyotrophic lateral sclerosis undergoing cesarean section

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To the Editor: Amyotrophic lateral sclerosis (ALS) is a relentlessly progressive neuromuscular disease associated with the degeneration of both upper and lower motor neurons. It eventually leads to muscle weakness, atrophy, and death within 3 to 5 years, most commonly from respiratory failure. The appropriate anesthetic management of these patients, especially parturients pose various challenges to anesthesiologists.

A 31-year-old woman at 37 weeks pregnant (65 kg, 165 cm) with progressive ALS was scheduled for cesarean section. She was diagnosed 13 months ago. She presented with dysarthria and it was difficult to lift her head away from bed. Motor power of her right and left proximal upper extremities were grade 3 and grade 4, respectively. She had mildly impaired swallowing and respiratory dysfunction, and she was requiring non-invasive ventilator at midnight, using bi level positive pressure (BiPAP) synchrony. Pre-operative examination revealed an amyotrophic lateral sclerosis functional rating scale revised (ALSFRS-R) score[1] of 28. Her family history showed remarkable hereditary feature. Her mother's brother was diagnosed with lower motor neuron syndrome. Her grandfather's brother presented neck weakness. They died 3 and 1 year after onset, respectively. She had delivered a boy at her age of 27 years. However, she experienced postpartum urinary retention as long as 40 days after vaginal delivery due to meperidine. Further pulmonary function test showed a restrictive pattern with low vital and total lung capacity (forced vital capacity: 52% of normal). Arterial blood gas analysis showed $PaCO_2$ of 34.1 mmHg and oxygen saturation of 98.7% with non-invasive ventilator.

After pre-operative multidisciplinary consultation, it was decided to implement bilateral ultrasound-guided transversus abdominis plane (TAP) block combined with incision infiltration before the delivery and to give total intravenous anesthesia (TIVA) with remifentanil and propofol by target-controlled infusion after the delivery to reach a proper analgesia.

Upon entry to the operating room, standard monitors were applied including 5-lead electrocardiogram with ST-segment analysis, non-invasive blood pressure, and pulse oximetry. Intra-operatively she was provided oxygen with her BiPAP ventilator. The patient was placed in the supine position and under ultrasound guidance, a 17 G Touhy needle was positioned via an in-plane approach at the level of mid-axillary line between sub-costal margin and iliac crest. Following this, 20 ml of 0.5% ropivacaine was injected through the needle with adequate spread between internal oblique and transversus abdominis in the left side, and another 20 ml in the right side [Figure 1]. Partial onset of sensory blockade was noted within 10 min and at the time of surgical incision, 0.5% lidocaine was injected around the incision. A live girl weighing 2650 g was born, with Apgar score of 10-10-10 (1-5–10 min). Anesthesia was maintained with 2 μg/mL (plasma target concentration, Marsh model) of propofol and 3 ng/mL (plasma target concentration, Minto model), which could relieve intra-operative discomfort. During the operation, the minimal oxygen saturation (SpO_2) was 98% and an arterial blood gas measurement shortly showed $PaCO_2$ of 31 mmHg and PaO_2 of 106 mmHg. The intensity of post-operative pain was less than 4 at 2 and 24 h on the numerical rating scale (NRS) without opioid treatment. She was discharged on postoperative day (POD) 3 without complications.

ALS is a degenerative disease of the motor ganglia in the anterior horn of the spinal cord and spinal pyramidal tracts. ALS remains the most common form of motor neuron disease with an incidence of 1.5 to 2.5 for 100,000 per year.[2] Autosomal-dominant familial cases account for nearly 10% of ALS cases. Recently mutations in the fused in sarcoma (FUS)/translocated in liposarcoma gene have been identified as a cause of familial ALS.[3]

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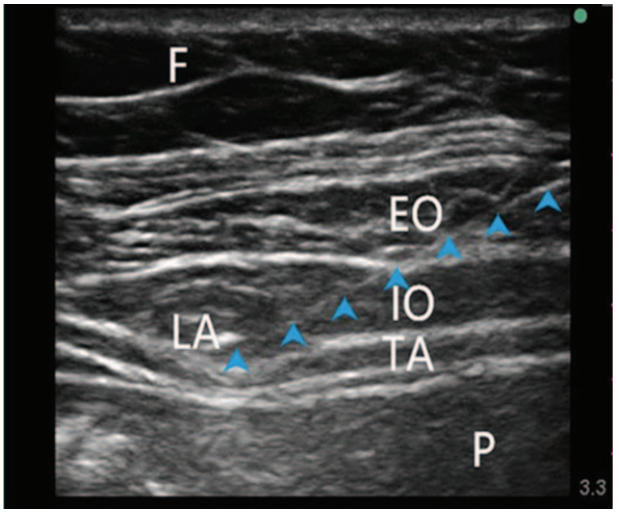

Figure 1: Sonographic anatomy of the US-guided TAP block. Images show the lateral abdominal wall using a probe held in the mid-axillary line in the axial plane. The right of the image is anterior. TAP: Transversus abdominis plane; EO: External oblique muscle; IO, Internal oblique muscle; TA: Transversus abdominis muscle; LA: Local anesthetic; F: Fat; P, Intraperitoneal structures; narrow arrow: needle with tip positioned in the TAP.

Her polymerase chain reaction analyses of total RNA revealed minor mutation on *FUS* (NM_004960) gene.

Cesarean section would be better rather than labor because of progressive weakness of respiratory muscles. Parturients with ALS may not meet respiratory demands during and after birth.

In choosing an anesthetic method for an ALS patient, anesthesiolgists should consider which method will be least harmful in terms of its progression. There is always the fear that the administration of the local anesthetic close to the nerve or needle trauma could exacerbate pre-existing disease symptoms.[4] Therefore, we avoided neuraxial block as a conservative and safe approach.

As pointed out general anesthesia (GA) has its risks. Patients with bulbar involvement can have post-operative aspiration and respiratory inadequancy. According to the literature,[1] the mortality rate may increase at ALSFRS-R scores less than 39 points. Therefore we chose peripheral nerve blockade to minimize the potential effects of local anesthetics on the spinal cord and TIVA without muscle relaxants for a smooth recovery after GA. Few other studies recommended the usage of ropivacaine in patients with ALS for peripheral nerve blockade. Belavy *et al*[5] used 0.5% ropivacaine (40 mL) for bilateral TAP blocks after cesarean section. It can block the level of skin, subcutaneous and muscle, but has no effect on visceral pain. Intravenous anesthetics can block visceral pain. Under sub-anesthetic concentrations of propofol, sevoflurane, remifentanil, and ketamine can decrease visceral pain-evoked potentials amplitudes. So we chose the rapid reversible short-acting analgesic (remifentanil) and amnestic (propofol) agents to relieve visceral pain. No muscle relaxant was applied in this parturient. Depolarizing neuromuscular blockers, such as succinylcholine should be strongly avoided because they can cause a lethal elevation of serum potassium in neuromuscular disorders like ALS. Non-depolarizing neuromuscular blockers should also be used sparingly and at the lowest possible doses with the use of neuromuscular function monitoring.

Through our article, we aim to bring out the fact that the use of a peripheral nerve blockade when possible could minimize the potential effects of local anesthetics on the spinal cord. And the overall strategy outlined includes the use of rapidly reversible short-acting analgesic and amnestic agents with no neuromuscular relaxants.

Declaration of patient consent

The authors certify that they have obtained the appropriate patient consent form. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflicts of interest

None.

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