Case Report
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Anesthesia management for ALS and WPW

Epidural anesthesia

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Abstract

Epidural anesthesia can provide anesthesia and analgesia for unilateral or bilateral lower extremity surgery and is associated with a low complication rate. We present our epidural anaesthetic management of a patient with both Amyotrophic lateral sclerosis (ALS) and Wolff-Parkinson-White (WPW) syndrome after intertrochanteric femur fracture surgery. It should be kept in mind that the choice of correct anaesthetic method in such patients with complicated neurological, pulmonary, and cardiac symptoms will significantly reduce postoperative mortality and morbidity.

Keywords

Epidural Anesthesia; Amyotrophic Lateral Sclerosis; Wolff-Parkinson-White Syndrome

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Introduction

Amyotrophic lateral sclerosis (ALS) is characterized by pathological degeneration of lower motor neurons, motor nuclei of the caudal brainstem, and descending pathways of upper motor neurons. Its clinical signs are bulbar paralysis with fasciculations and progressive muscle atrophy. Since the disease often involves atrophy and weakness of respiratory muscles resulting in respiratory failure and death, anaesthetic management of patients with ALS has been a controversial topic [1].

Wolff-Parkinson-White (WPW) syndrome is an uncommon cardiac condition where there is an abnormal band of atrial tissue connecting atria and ventricles, which in turn can electrically bypass the atrioventricular node. Anaesthetic management in these patients is extremely difficult due to probable life threatening complications such as paroxysmal supraventricular tachycardia and atrial fibrillation which may occur perioperatively [2]. We report our successful anaesthetic management of a patient with both ALS and WPW syndromes.

Written informed consent was obtained for this report.

Case Report

The patient is a 59-year-old, 97 kg, 1.80 m male patient with a 3-year ALS history, who was scheduled for intramedullary nail (IMN) for the right femur fracture. The patient also suffered from obstructive sleep apnea syndrome (OSAS), chronic obstructive pulmonary disease (COPD), type 2 diabetes mellitus (DM), and WPW diseases. ECG was compatible with WPW (Figure 1. Short PR and delta wave). He does not use CPAP at home. During physical examination, the patient was completely conscious and cooperative. There was no active movement in the lower limbs and muscle strength was 2/5. Muscle strength in the upper extremities was 3/5. He could sit for short periods of time by himself. He could walk with aid. He had difficulty speaking and swallowing and had fasciculations. His Mallampati score was 3 according to airway evaluation. Spontaneous respiratory effort was sufficient, and respiratory voices were slightly coarse on the bases of osculation. He was evaluated as American Society of Anesthesiologists (ASA) III. During the preoperative visit, he said that he did not want general anesthesia and demanded regional anesthesia if possible.

The patient was taken to the operation room and infusion of 0.9% NaCl was started. For premedication, 2 mg midazolam was given intravenously. The left radial artery was cannulated. ECG, pulse oximetry, invasive arterial blood pressure, and body temperature were monitorized. Preoperative heart rate was 90/min, blood pressure was 110/50 mmHg, respiratory rate was 15/min, and oxygen saturation was 97%. The preoperative arterial blood gas values were pH 7.38, PCO2 31.8 mmHg, PO2 80.1 mmHg, SO2 96.2%, and HCO3 22.4 mmol/L. After 15 minutes, an epidural catheter was inserted through the third lumbar vertebral interspace, using a 17-gauge Tuohy needle, while the patient was in the left lateral decubitis position. After the catheter was taped to the skin, the patient was returned to the supine position. The patient was given a humidified mixture of air and oxygen at 2 L/min through a nasal cannule. A test dose of 2 ml of 2% lidocaine was injected through the epidural catheter. The patient was hemodynamically stable. Five minutes later, an additional 5 ml of 2% lidocaine and 5 ml of 0.5% bupivacain were injected through the catheter. After 20 minutes the sensory block level, which was measured by pinprick test, was at T8 level. Additional 2% lidocaine 3 ml and 0.5% bupivakain 3 ml were injected through the catheter for analgesia at the 90th minute. The intraoperative hemodynamic was stable (blood pressure ranged from 90/70 to 120/90 mm Hg, heart rate 80 to 100 beats/minute without arrythmias). The intraoperative arterial blood gas values were pH 7.44, PCO2 35.8 mmHg, PO2 87.3 mmHg, SO2 97.6%, and HCO3 25.4 mmol/L. Total blood loss was 300 ml. In total, 1000 ml 0.9% isotonic NaCl and 400 ml colloid solutions were transmitted. The surgery was completed with no complications and lasted two hours. No respiratory complication was experienced during the operation. The patient stayed in PACU for one hour after surgery. During the PACU stay he was stable. There was no nausea or vomiting. The VAS score on PACU arrival was 0/10. Vital signs were as follows: heart rate 104, blood pressure 110/78, respiratory rate 19, and oxygen saturation of 97% on 2 litres nasal cannula. After a stable PACU stay, the patient was observed at the orthopedics clinic. Motor block level was evaluated with the Modified Bromage scale (scale 0=full flexion of foot, knee, and hip, i.e., no motor block; scale 1=full flexion of foot and knee, unable to perform hip flexion; scale 2=full flexion of foot, unable to perform knee and hip flexion; and scale 3=total motor block, unable to perform foot, knee, and hip flexion). Motor function recovery of the lower extremities to the preoperative level was completed within two hours postoperatively. Patient-controlled analgesia device was connected with 0.1% bupivacaine in 0.9% isotonic solution through the epidural catheter for postoperative analgesia. There was no infusion dose, bolus dose was 10 ml, and locked time was 20 minutes on the PCA device. The patient was advised to press the button of the PCA device when he felt pain. In short, the postoperative period was stable, uneventful, and successful, and the patient experienced almost no incisional pain. The patient was discharged 10 days after surgery.

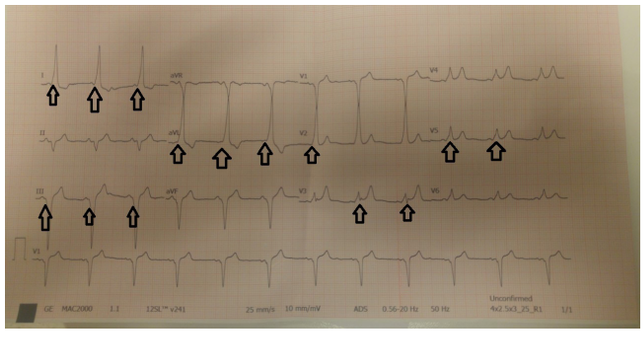

Figure 1.

Discussion

ALS is a progressive disease with unknown etiology characterized by motor neuron degeneration in the cerebral cortex, brain stem, and spinal cord. ALS does not affect the respiratory system directly but it damages mechanical function of the respiratory system by affecting expiratory and inspiratory muscles and the upper airway muscles. Thus, respiratory involvement in ALS patients is one of the major reasons of death [1]. After denervation and prolonged immobilization, upregulation of acetylcholine receptors occur at the neuromuscular junction and along the skeletal muscle membranes. Depolarizing neuromuscular blockers, such as succinylcholine, can lead to activation of an unexpected large quantity of receptors resulting in an abnormally high efflux of potassium. The possibility of a sudden increase in plasma potassium levels and resultant ventricular arrhythmia or fibrillations after the usage of succinylcholine in these patients has also been reported by several investigators [3]. For this reason we did not use succinylcholine.

Patients with ALS are highly sensitive to nondepolarizing muscle relaxants, as are patients with myasthenia gravis. Extubation may be difficult as a result of baseline muscle weakness and altered pulmonary function. Intraoperative anaesthetic management of these patients should include the use of rapid, reversible, short-acting anaesthetic and analgesic agents and to avoid (depolarizing) and/or minimize (nondepolarizing) muscle paralysis.

Exacerbation of pre-existing neurological damage after spinal and epidural anesthesia has been described by Kane [4] and Vandam and Dripps [5]. However, most of the reported cases were related to coexisting lumbar disc hernia, spinal tumor, or technical problems. To the best of our knowledge, there is no data about neurological dysfunction of the spinal cord in patients with neurodegenerative disease after epidural anesthesia. This does not mean that epidural anesthesia is always secure for patients with ALS, but we believe that the risk of general anesthesia is greater than a well-managed epidural anesthesia. It should not be forgotten that lumbar epidural anesthesia may cause respiratory depression if the sensory block level is above T6, since the expiratory reserve volume can be reduced. In our case, the sensory block level was T8. Epidural anesthesia is safer than spinal anethesia for better hemodynamic stability.

Wolff-Parkinson-White (WPW) syndrome is a ventricular preexcitation syndrome resulting from aberrant conduction pathway. The incidence of WPW syndrome is 0.9–3% and the risk of sudden death due to a malignant arrhythmia is estimated at 0.4% yearly in these patients. The aim of anaesthetic management should be the avoidance of sympathetic stimulation such as pain, anxiety, and stress response to intubation and hypovolaemia. Regional anesthesia is preferred over general anesthesia in order to avoid multidrug administration [2]. Blockade of cardiac accelerator fibers and suppression of normal AV conduction might occur at high subarachnoid block. Therefore, we performed epidural anesthesia, which allows the drug dose to be titrated slowly.

Although there are case reports with either ALS or WPW syndromes in literature, we did not observe any case reports in which the two syndromes were both present as in our case. Hara et al. performed epidural anesthesia on a male patient aged 69 years with ALS who underwent a right inguinal hernia repair operation and reported no complications [1]. Anis and Anthony performed thoracic paravertebral block with multimodal anesthesia on a 64-year-old patient diagnosed with ductal carcinoma with Primary Lateral Sclerosis (PLS) who underwent axillary lymph node dissection and mastectomy, and they reported no complications [6]. Pravalika and Viyanak performed spinal anesthesia on a 20-year-old female patient with WPW in order to terminate a 13-week molar pregnancy and curettage; they also reported no complications [7].

In this case, peripheral nerve block techniques (lumbar plexus and sciatic nerve block) could be preferred for anesthesia management [8]. We didn't select this technique because of the disadvantages such as long-term motor blockade and high-dose local anaesthetic. In addition, epidural catheterization provided better postoperative analgesia management and for long-term rehabilitation.

Continuous spinal anesthesia (CSA) is an another technique which can be preferred, because this technique provides well balanced hemodynamic stability and anesthesia [9]. But we didn't perform CSA since our postoperative analgesia team does not have enough experience in CSA catheter treatment during the postoperative period.

Regional anesthesia techniques can be successfully used to prevent respiratory and other complications that may be associated with general anesthesia and opioid use. In our case, there were no complications such as respiratory, motor function loss, or cardiac arrhythmias in the postoperative period, and this can be evaluated as a success for our practice.

Conclusion

We reported our epidural anesthesia practice on a patient with comorbidities including ALS, WPW, COPD, and OSAS. We believe that the correct choice of anaesthetic method in such patients with complicated neurological, pulmonary, and cardiac symptoms will significantly reduce postoperative mortality and morbidity.

Scientific Responsibility Statement

The authors declare that they are responsible for the article's scientific content including study design, data collection, analysis and interpretation, writing, some of the main line, or all of the preparation and scientific review of the contents and approval of the final version of the article.

Animal and human rights statement

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. No animal or human studies were carried out by the authors for this article.

Conflict of interest

None of the authors received any type of financial support that could be considered potential conflict of interest regarding the manuscript or its submission.

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