Anesthetic management of a patient with amyotrophic lateral sclerosis for transurethral resection of bladder tumor

INTRODUCTION

Amyotrophic lateral sclerosis (ALS), a progressive neurological disease with incidence of 1:50,000,[1] is characterized by degeneration of upper and lower motor neurons due to degeneration of lateral corticospinal tracts in the spinal cord and consequent muscle denervation. Eventual respiratory muscle failure makes it fatal.

CASE REPORT

We report anesthetic management of a patient with ALS who presented for transurethral resection of the bladder tumor (TURBT).

A 45-year-old male presented with a history of hematuria for 2 weeks. Cystoscopy revealed papillary growth 1 × 0.5 cm on the left side of bladder base, extending to the left wall. He was diagnosed to have ALS four years ago, when he had developed a slow onset, progressive, asymmetric weakness of upper limbs and lower limbs with associated difficulty in walking. He was a non-smoker with no other co-morbid illnesses. Neurologically, he had spasticity of upper and lower limbs with exaggerated tendon reflexes. His motor power was grade 3 (MRC grading)[2] in upper limbs and grade 4 powers in lower limbs [Table 1] and associated hand muscle weakness with widespread fasciculations. He was bradykinetic with resting tremors of all extremities. There was no evidence of bulbar dysfunction or truncal weakness.

Preoperative examination demonstrated ineffectual cough, with borderline peak expiratory flow rate (450 L/min), and reduced breath sounds in both lung bases. Further pulmonary function test showed a restrictive pattern with low vital and total lung capacity with preserved flow rates (FVC: 60% of normal; FeV1: 58% of normal; FEV1/FVC: 96%). Blood investigations including serum electrolytes were normal (s.Na^+: 134 mmol/L; s.K^+: 4.9 mmol/L; s. HCO_3 –20 mmol/L).

His regular medications which included Syndopa, Ropinirole, Clonazepam, Amitriptyline and Trihexyphenidyl were continued till the morning of surgery.

A combination of general anaesthesia without muscle paralysis and a regional block was planned for surgery. Informed consent was obtained prior to the procedure

Table 1: Medical research council grading of motor power[2]

Grade 5	Normal power
Grade 4	Active movement against gravity with resistance
Grade 3	Active movement against gravity without resistance
Grade 2	Active movement with gravity eliminated
Grade 1	Only a trace or flicker of movement
Grade 0	No movement

and he was fasting to solids for 6 h and to water for 2 h. No premedication was given.

General anaesthesia was induced with propofol 2 mg/kg and fentanyl at 1 mcg/kg via a 20-gauge IV cannula. Ventilation was assisted with a mixture of air–oxygen and isoflurane. Ensuring adequate depth of anaesthesia, airway was secured with size 4 laryngeal mask airway (LMA). Anaesthesia was maintained on 50-50 mixture of air–oxygen with isoflurane on spontaneous respiration.

Bilateral obturator block with 15 mL of 0.5% hyperbaric bupivacaine on each side was performed after LMA insertion. A nerve stimulator was used for eliciting adductor response between current of 0.4-0.6 mA. No drug interactions were observed. Surgery lasted for one hour, during which he received 400 mL of Ringer lactate solution. At end of the procedure, patient was extubated awake. He had an uneventful recovery in the post-operative period and was discharged on the third post-op day.

DISCUSSION

The spectrum of amyotropic lateral sclerosis' involvement of skeletal muscles raises concerns regarding the technique of anaesthesia.

Respiratory involvement, though a less common pattern of ALS onset, includes respiratory muscle weakness (1 to 3%),[3] generalized weakness in the limbs and bulbar muscles (1 to 9%), axial onset with head drop or truncal weakness. Patients with respiratory muscle weakness initially note fatigue, progressing to shortness of breath triggered by decreasing levels of activity and lying flat. They develop disturbed nocturnal sleep with frequent awakenings and excessive daytime sleepiness. The Borg dyspnea scale, a non-invasive test, has been used as a simple marker to predict respiratory muscle weakness in ALS.[4]

Loss of innervation ultimately leads to muscle atrophy with extra-junctional and hypersensitive nicotinic acetylcholine receptors. General anaesthesia with use of muscle relaxants cause ventilatory depression due to abnormal responses to muscle relaxants.[5] Depolarizing muscle relaxants elicit neuromyotonia-like contractions, rhabdomyolysis and severe hyperkalemia.[6] Non-depolarizing muscle relaxants can be administered but sensitivity to these drugs is altered as in upper and lower motor neuron lesions, immobilization and burns. Respiratory dysfunction and pathologic reaction to muscle relaxants lead to anesthetic complications.

The use of regional central neuraxial block is also relatively contraindicated for fear of exacerbating progression of disease pattern.[7] Neuraxial anaesthesia may cause relapses or exacerbation of the disease as a result of needle trauma, technical difficulties, drug toxicity or the selection of certain substances such as vasopressors or lidocaine.[8] The mechanism by which neuraxial anaesthesia may exacerbate the disease is unknown. Diagnostic lumbar puncture alone does not seem to be associated with deterioration of symptoms. The lack of a protective nerve sheath around the spinal cord and associated demyelination may render the spinal cord more susceptible to potential neurotoxic effects of local anesthetics.[9] Because local anesthetic concentrations are significantly smaller within the white matter of the spinal cord after epidural administration, this modality of neuraxial anaesthesia is generally recommended over intrathecal techniques.[10]

Our patient had a tumor of the bladder involving lateral wall, raising the requirement of motor block. We decided against the use of muscle relaxants in view of its obvious dangers. Therefore, a general anesthetic technique with the patient on spontaneous ventilation and airway secured by LMA was decided on, since our patient had no evidence of bulbar dysfunction or truncal weakness. In view of conflicting reports with central neuraxial block and no definite literature regarding use of regional nerve blocks, obturator block was considered with aid of a nerve stimulator to decrease incidence of direct nerve injury.[11] Epidural technique was not preferred because of the risk of having an incomplete/patchy motor block. Spinal anaesthesia can cause direct stimulation of obturator nerve, whereas general anaesthesia with nerve block avoids this.

CONCLUSION

Patients with ALS may need surgical intervention for concomitant problems. The goal of anaesthesia would be choosing a technique that least interferes with disease pattern, has no deleterious effect on the patient and yet provides optimum operating conditions and adequate analgesia. The individual patient characteristics, type of surgery, and above all, risk versus benefit of the technique considered need to

be evaluated before choosing to anesthetize a patient with ALS. A non-paralytic general anaesthesia with regional nerve block ensured anesthesia, immobility and analgesia in our patient.

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