Scholars Journal of Applied Medical Sciences (SJAMS)
Abbreviated Key Title: Sch. J. App. Med. Sci.
ISSN 2320-6691 (Online)
ISSN 2347-954X (Print)

A Unit of Scholars Academic and Scientific Society, India
www.saspublishers.com

Anaesthesiology

Anaesthetic Consideration for Caesarean Delivery in a Parturient With Motor Neuron Disease

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Case Report

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Article History
Received: 09.12.2017
Accepted: 12.12.2017
Published: 30.12.2017

DOI:
10.36347/sjams.2017.v05i12.040

Abstract: Parturient with pre-existing central nervous system disorder like motor neuron disease poses a unique challenge to the anaesthesiologist. The choice of proper anaesthetic technique is still contentious and depends on patient's condition at the time of presentation. We describe the anaesthetic management of a patient with motor neuron disease presented for emergency caesarean delivery.
Keywords: Motor Neuron Disease, Amyotrophic lateral sclerosis, Pregnancy, Emergency Caesarean Delivery, General anaesthesia, i-gel supraglottic airway device.

INTRODUCTION

Motor Neuron Disease(MND) is a devastating and progressive neurodegenerative condition of undetermined aetiology characterised by loss of upper and lower motor neurons in the spinal cord, cranial nerve nuclei and motor cortex resulting in denervation of the skeletal muscles leading to spasticity or atrophy depending on the location of the abnormality. Amyotrophic Lateral Sclerosis (ALS) or Lou Gehrig's disease is the most common form of progressive motor neuron disease which involves the anterior horn cells. The characteristic features of ALS include loss of all voluntary motor function, with the exception of the ocular muscles and sphincters. Sensation and higher cortical function are also unaffected. Death usually results from compromise of the respiratory muscles within 5-6 years [1].

MND/ALS in women of reproductive age is extremely rare [2]. It is more common in male patients in their 50s and 60s with an increase with age until the seventh decade.

Hospital based studies from India have reported higher male to female ratio of 2:1 to 7:1. Most of the available data suggests earlier onset of ALS in Indian population by one to two decades. The younger population have greater neuronal reserve which is possibly the reason for the longer survival in the Indian population [3].

CASE HISTORY

A 27-year-old primigravida, nulliparous female patient weighing 54 kg and height of 162 cm with 37 weeks of gestation was posted for emergency lower segment caesarean section (LSCS) for foetal distress (meconium stained amniotic fluid). She was diagnosed to have ALS under El Escorial World Federation of Neurology criteria [4] by consultant neurologist two years earlier & was on Tab Riluzole 100 mg/day for last 14 months. She also had history of fall from two-wheeler six months prior to the diagnosis of MND with complain of pain in the right lower limb and was treated symptomatically. But after six months of injury, she had history of fever following which she developed difficulty in walking with pain in left lower limb, difficulty in wearing slippers. Since the last six months, her weakness progressively spread to both upper limbs (Left>Right) resulted in difficulty in wearing clothes and combing hair. She had no history of headache, urinary incontinence, vomiting, seizures or any other co-morbid illness. There was no contributory family history. On examination, her higher function test, speech, cranial nerves and sensory system appeared to be normal. Motor system examination revealed spasticity of upper & lower limbs with brisk Deep Tendon Reflexes (grade 3/5) and diminished power (MRC grade 3) [Table 1].

There was wasting of thenar and hypothenar muscles of both hands (Left>Right) and bilateral foot

drop (Left>Right). Superficial reflexes were within normal limits except extensor planter reflex (positive Babinski sign). There was no evidence of bulbar dysfunction or truncal weakness. EMG & NCV study were suggestive of anterior horn cell disease as evidenced by positive sharp waves in both upper & lower limb muscles studies, recruitment of motor unit potentials was reduced and decreased nerve conduction velocity mainly in the lower limb nerves. MRI(1.5 T) of lumbo-sacral spine revealed straightening of lumber spine, mild diffuse disc bulge at level of L_{4-5} and L_5-S_1, narrowing the both exit neural foramina; mild facet joint hypertrophy noted at L_5-S_1 level with normal screening of cervical and thoracic spine. The patient was bedridden because of weakness and low molecular weight heparin 40 mg daily was given as prophylaxis to prevent formation of deep vein thrombosis.

Pre-anaesthetic airway assessment revealed Mallampati grade II, adequate flexion & extension of neck with thyromental distance of 6.5 cm. Baseline heart rate was 96 beats per minute, BP 146/80 mm Hg, O_2 saturation of 97% on room air. She was complaining of slight shortness of breath (Modified Borg Dyspnoea Scale score 2). [Table 2] Cardiovascular and GI systems were normal on clinical examination. Respiratory examination revealed effective cough reflex and adequate air entry in both lungs. Pulmonary function test (PFT) showed mild restrictive pattern. Her complete blood count, serum biochemistry, liver function tests, coagulation profile, thyroid function tests and urine examination reports were within normal limits. A diagnosis of ASA grade IIIE was made. High risk written & informed consent was obtained. General Anaesthesia (GA) with muscle relaxant Supraglottic airway device (SAD) insertion and assisted ventilation was planned in view of foetal distress, h/o trauma to the lumber spine, MRI findings and risk of exacerbation of neurological findings.

On arrival in the operating room, one intravenous line was secured with 16G cannula. Standard monitors (NIBP, pulse oximeter, ECG) were attached. Aspiration prophylaxis with inj. Metoclopramide (10mg) and inj. Ranitidine (50mg) was given. She was premedicated with intravenous inj. Glycopyrrolate (0.2mg) & pre-oxygenated with 100% oxygen for 3 minutes. Surgeons were asked to paint and drape the surgical field, and then induction was accomplished by using sellick's manoeuvre with inj. Propofol 100 mg and inj. Fentanyl 50 µg. Once the parturient became unconscious, a size 4 i-gel SAD was inserted at the first attempt after lubricating the device with a water-based lubricant. After insertion, i-gel was fixed by adhesive tape and adequate ventilation was confirmed by clinical observation of chest wall movement, bilateral chest auscultation, listening to escape of gas from the mouth, presence of square wave of capnogram and lack of gastric insufflation. Cricoid pressure was then released and a size 12 FG gastric tube was inserted through gastric channel. Decompression of patient's stomach was done and the gastric tube was connected to collection bag to drain freely during surgery.

Anaesthesia was maintained with oxygen, N_2O, sevoflurane (1.5-2%) and assisted ventilation. A healthy baby girl weighing 2.7 kg was delivered with an apgar score of 7 and 9 at one and five min respectively. After the delivery, 5 unites of oxytocine was given as bolus IV over 3 min and then 10 unites of oxytocin was added to 500 ml of ringer lactate and Inj. Fentanyl (1 µg/kg) IV bolus was administered. The duration of the surgery was 35 min and her vitals remained within normal range throughout this period. At the end of surgery, 1 gm inj. Paracetamol was given intravenously and bilateral USG guided transversus abdominis plane (TAP) block was performed with 0.375% Ropivacaine (0.6 ml/kg with half the volume on either side) for postoperative analgesia.

The i-gel airway device was removed after thorough oral suction, once the patient had regained full consciousness and was generating adequate spontaneous ventilation. She had mild cough immediately after extubation which was subsided within few minutes. She was shifted to the recovery room with oxygen at 5 L/min via simple face mask and 45 º head-up positions. Inj. aqueous diclofenac sodium 75 mg intravenously was also used as a part of multimodal analgesia. The patient was under observation in our recovery room for 4 hours and in ward for next 7 days and there was no neurological deterioration.

Table-1: Medical Research Council (MRC) Grading of muscle power

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

Table-2: Modified Borg Dyspnoea Scale

Score	Shortness of breath
0	Nothing at all
0.5	Very, very slight (just noticeable)
1	Very slight
2	Slight
3	Moderate Exercise Training Zone
4	Somewhat severe
5	Severe
6	
7	Very severe
8	
9	Very, very severe (almost maximal)
10	Maximal

DISCUSSIONS

The published literature on anaesthetic management for caesarean delivery of parturient with ALS is exiguous. The largest experience of pregnancy with ALS is probably in Guam in 1956(detailed 21 pregnancies in 17 women), where the incidence is approximately 100 times greater than elsewhere. No effect, either beneficial or detrimental, of ALS on pregnancy (or vice versa), has been noted from Guam [5]**.**

The main concern for the anaesthesiologist in patient with ALS is involvement of upper airway and respiratory muscles including reduced vital capacity. Poor cough and bulbar dysfunction lead to aspiration. In pregnancy, there is 45% increase in the resting minute ventilation, resulting mainly from a rise in tidal volume as respiratory rate remains unchanged [6]**.** Also late in pregnancy, the diaphragmatic elevation caused by enlarged uterus leads to 20% decrease in functional residual capacity. Thus, Patient's respiratory compromise should be assessed and serial follow up during pregnancy & throughout the peripartum period is recommended. The Borg dyspnoea scale is a valuable non-invasive test for the prediction of inspiratory muscle weakness in ALS patients [7]**.** However despite this theoretical risk of increased respiratory complications, most described cases in literature were not adversely affected and neither was the course of the disease.

Choice of anaesthetic technique for LSCS in patient with ALS, based on pulmonary function alone did not favour either regional or general anaesthesia convincingly. The proper anaesthetic technique is controversial and there is no precise guideline. Although central neuraxial block is not absolutely contraindicated, there is fear of exacerbation of pre-existing disease pattern [8]. The worsening of neurologic outcomes are secondary to an increased risk of neurologic injury from needle or catheter-induced mechanical trauma, local anaesthetic toxicity as a result of increase susceptibility due to demyelination, neural ischemia secondary to local anaesthetic additives, personal biases from the patient and potential medicolegal implications[9]**.** Epidural anaesthesia has been administered for LSCS in patient with ALS without evidence of worsening of neurologic function postoperatively [2].

The use of muscle relaxants during GA is problematic in patients with pre-existing neuromuscular disorders. Use of depolarizing muscle relaxant succinylcholine is contraindicated because of the high risk of life-threatening hyperkalemia which may result in ventricular fibrillation or asystole and rhabdomyolysis [10]**.** Circulatory collapse after administering succinylcholine in a patient with diffuse motor neuron disorder has been reported [11]. Nondepolarizing muscle relaxants should be used with caution because of a possible exaggerated response due to increased sensitivity. Jang-hyun Kim *et al*. administered general anaesthesia in a patient with Kennedy's disease for laparoscopic cholecystectomy. They had monitored the degree of neuromuscular blockade throughout the operation and injected atracurium in one third of usual intubating dose for induction and one fifth of the induction dose on demand for maintenance.

The ET tube was removed 16 hours after surgery because of delayed recovery but there was no exacerbation of neurologic signs or symptoms postoperatively [12]. The study of Kim *et al.* indicates that a low dose of rocuronium appears feasible and safe in an ALS patient [13]. We decided to avoid muscle relaxant in order to avoid the possibility of ventilatory depression due to abnormal responses to muscle relaxants, exacerbation of the motor neuron disease [14] and due to unavailability of neuromuscular monitoring and sugamadex in our theatre. Amin *et al.* had compared the efficacy of i-gel and endotracheal tube on patients undergoing elective caesarean section and concluded that i-gel can replace the need of tracheal intubation in elective LSCS without any serious complications as compared to tracheal intubation [15]. I-gel SAD for airway maintenance has also been used for emergency LSCS in a patient with kyphoscoliosis

[16]. We also preferred to use i-gel SAD as high seal pressure is required in pregnant patients and a ProSeal laryngeal mask airway was not available in theatre.

Inhaled anaesthetics not only potentiate the action of neuromuscular blocking agents but also have some intrinsic muscle relaxants properties. Sevoflurane produce skeletal muscle relaxation that is about twice as high as that associated with halothane [17]. We used N_2O and sevoflurane for maintenance. These agents provided a good operating condition for the surgeon without the use of any muscle relaxant. We used bilateral TAP block in our patient as a part of multimodal pain management without any deterioration of neurological condition. Thampi *et al.* administered non-paralytic GA with obturator nerve block in a male patient with ALS undergoing transurethral resection of bladder tumour without any complication [18].

CONCLUSION

We have successfully administered non-paralytic general anaesthesia with i-gel supraglottic airway device and TAP block in a parturient with motor neuron disease for caesarean delivery to provide optimal surgical condition and adequate analgesia without any neurological deterioration.

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