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Anaesthetic Management of Parturient with Motor Neuron Disease for Caesarean Delivery: Case Report

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ABSTRACT

Motor neuron disease (MND) is a progressive neurodegenerative disease of unknown aetiology, which results in weakness of muscles of phonation, ambulation, deglutition and respiration. It has low prevalence but high disability and fatality. Death often follows respiratory failure. There is no known cure. It is extremely rare in pregnancy, but when it occurs, the respiratory compromise worsens and anaesthesia becomes challenging. There is no consensus yet on the choice of anaesthesia. We present a 29-year-old, 32-week parturient with MND who presented with severe dyspnea, orthopnea and was diagnosed with severe respiratory distress. She was admitted into ICU and given anticoagulant and steroid therapy. Sequential mini-dose combined spinal-epidural anaesthesia (CSE) was given and surgery performed in the semi-sitting position successfully. There was no deterioration of symptoms post anaesthesia. We conclude that CSE is adequate to manage parturient with MND who has severe orthopnea following respiratory muscle paresis.

Keywords

Motor neuron disease, Parturient, Combined spinal-epidural, Anaesthesia.

Key Message

CSE is good for management of MND in pregnancy. Where severe respiratory distress and orthopnea are major complaints, sequentially topping up epidural after a mini-dose spinal is useful.

There is no evidence of worsening neuromuscular function after use of regional anaesthesia in parturient with MND.

Introduction

Motor neuron diseases (MNDs) are progressive neurodegenerative disorders that weaken muscles. The weakness affects muscles of phonation, ambulation, deglutition and respiration. The neurons affected may be upper or lower motor neurons or both (as in amyotrophic lateral sclerosis). A disease of unknown aetiology, genetic, viral and inflammatory cause has been suggested [1]. MNDs have low prevalence and incidence but high disability and fatality rate [2] and are commoner in male than female with preponderance in fifth and sixth decade [3]. Fatality occurs within 3-5 years with some living to 10 years. Death often follows respiratory failure. There is no known cure.

MND is extremely rare in pregnancy. When MND patient becomes pregnant, respiratory changes during pregnancy are exaggerated. The diaphragm is the major muscle of respiration. During advanced pregnancy, the uterus splints the diaphragm and with the intercostals weakened by MND, respiratory insufficiency develops. The resulting dyspnea often requires Caesarean section. Anaesthesia for patients with MNDs is challenging because of respiratory compromise. There is no consensus on the best anaesthesia. The use of suxamethonium is contraindicated due to risk of hyperkalemia and sensitivity to non-depolarisers is heightened often-requiring prolonged post anaesthesia ventilatory support [4].

This case reports successful management of parturient with MND for caesarean section using combined spinal-epidural anaesthesia.

This work has been reported in line with the SCARE criteria [5].

Case Presentation

AA, 29-year-old para 3, 32-week pregnant Saudi housewife, weighing 100kg (BMI 32.3kg/m^2) with three previous sections was brought in an ambulance from Neurologists to the Emergency Unit for repeat caesarean section due to severe dyspnoea from motor neuron disease (lower motor type). Patient was diagnosed three years earlier and managed with tab Riluzole 50mg BD. The previous sections occurred before MND's onset.

Dyspnoea started three months before presentation to Neurologists. It was progressive, and worsened over five days. It was associated with severe orthopnea. There was weakness of the upper and lower limbs, difficulty in phonation and swallowing along with non-productive cough. An only wife of a businessman, her father and uncle respectively died of MND 15 and seven years earlier. No history of alcohol or cigarette use. System review was essentially normal.

Physical examination revealed pregnant female propped in semi-sitting position by pillows on bed adjusted to 60º, with flaring alae nasi and supra-sternal recession. She was on oxygen via nasal prongs. Mildly pale, anicteric with bilateral pitting angular pedal oedema, respiratory rate was 36 cycles/minute. Breath sound was normal on both apices but inaudible bilaterally on middle and basal zones. Oxygen saturation was 94% (2L/min O2) with pulse rate 142/min, blood pressure 124/56mmHg and heart sound I and II only. She was conscious, alert, well-oriented in time, person and place. Sensation to touch and pain were normal globally. Power was 3/5 on lower limbs and 2/5 on upper limbs. Transverse suprapubic scar and 32-week-sized uterus were revealed. The lumbar spine was palpable dorsally.

Investigations revealed sinus tachycardia on ECG. Electrolytes, urea and creatinine were normal. Urinalysis was normal. Haemoglobin 10.9g/dl (11.5-16.6), hematocrit 34.9% (35-45) and platelet was 222 (140-400x103/dl). Coagulation profile was normal. Chest radiograph was not done due to obstetricians' concerns. Table below shows arterial blood gas (ABG) results.

A diagnosis of severe maternal respiratory distress secondary to progressive MND with three previous caesarean sections was made. A differential diagnosis of pulmonary embolism was entertained and anticoagulation therapy initiated with intravenous enoxaparin 4000IU (Clexane®, Sanofi-Aventis, Arabia) 12hourly. IV dexamethasone 8mg (Eipico, Egypt) daily was given to mature fetal lungs. She was transferred to intensive care unit (ICU) for monitoring.

At the ICU, patient was managed in the propped up position with oxygen entrained by nasal prong (2l/min). Daily ABG analysis was ordered. Due to severe restriction of respiratory efforts and worsening acidosis, the decision was made to operate at 33 weeks. Anticoagulant was stopped (regional anaesthesia precaution).

At pre-anaesthetic evaluation, patient's forced vital capacity was 200ml using peak flow meter (Breathing Exerciser®, Jam Joom Medical Industries Ltd., Saudi Arabia) against a predicted range of 2500ml. Blood pressure was 135/55mmHg, heart rate 103beats/min and SpO_2 was 96% on oxygen. A decision for combined spinal-epidural anaesthesia was taken and case conference conducted with the husband and high-risk consent obtained. IV metoclopramide 10mg (Premosan®, Gulf Pharmaceutical Ind., UAE) and ranitidine 50mg (Rantag®, Gulf Pharmaceutical Ind., UAE) were given. Patient was transported to the operating room (OR) propped in semi-sitting position. The planned anaesthesia technique was discussed with the Consultant Obstetricians. Patient would be in semi-sitting position for surgery.

In the OR, preparation was made for any emergent awake intubation; Glidescope® video laryngoscope (Verathon Medical, Canada), lidocaine 2% spray, endotracheal tubes and laryngeal mask airways were prepped. A 500ml of normal saline infusion (Pharmaceutical Solution Industry, Saudi Arabia) was set up. A sterile table onto which the contents of epidural set (Saudi Mais Medical Products Co., Saudi Arabia) was prepared. In the sitting position, the skin of the lower back was prepped with povidone iodine 10% solution (Alphadine®, Riyadh Pharma Ltd., Saudi Arabia). A size 16G Tuohy needle (150mm length) was passed at the L3/4 interspace and the epidural space identified using the loss of resistance to saline. A size 25Gx6" pencil point spinal needle (Saudi Mais Medical Products Co., Saudi Arabia) was passed into the L4/5 interspace and the intrathecal (IT) space entered

Parameter	At Presentation	Reference values [9]	Interpretation	Day 5 in ICU	Interpretation
pH	7.372	7.36–7.44	Low Normal	7.349	Acidosis
pCO^2	48.9	35-45mmHg	High (Respiratory Acidosis)	53.9	Worsening Respiratory acidosis
pO2	56.5	80-100mmHg	Low (Hypoxemia)	153	Oxygen therapy
HCO^{3-}	26.4	22-26mmol/L	Slightly high	26.9	Renal compensation
Base Excess	3.0	+2 to -2	compensating	3.7	Compensating
sO_2*	88.5	95-100%	Hypoxemia	97.9	Improved by oxygenation

Table showing ABG results at presentation and fifth day admission.
*sO_2 = arterial oxygen saturation, pO_2 = partial pressure of oxygen, pCO_2 = partial pressure carbon dioxide.

as confirmed by flow back of clear cerebrospinal fluid. Heavy 0.5% bupivacaine 7.5mg (Marcaine®, AstraZeneca, Switzerland) premixed with 15mcg of fentanyl (Martindale Pharmaceuticals, UK) was administered IT. The spinal needle was withdrawn. An epidural catheter was then introduced into the Tuohy needle and 5cm of catheter threaded into the epidural space. No blood or CSF flowed back. The catheter was taped in place. The operating table head was wound up to 60º and she rested on it. Patient was still dyspneic until a pillow was placed behind further propping her forward. A saddle block with loss of pain sensation up to the umbilicus (T10) was obtained without any change in vital signs. Surgery was allowed to proceed. Male baby weighing 1980g, with Apgar score 9, 10 was delivered with patient in semi-sitting position. During the exteriorization of the uterus, patient felt pain. The epidural catheter was then loaded with 5ml of 0.25% plain bupivacaine (Bucaine®, Hikma Pharmaceuticals, Jordan). Surgery continued. After this, blood pressure dropped below 20% of the pre-induction value. Aliquots of IV ephedrine 6mg (Martindale Pharmaceuticals, UK) up to 30mg were given and 250ml of Human Plasma Protein Solution 5% (Octapharma AG, Switzerland) set up. BP normalized. Epidural dose (5ml) premixed with 0.2mg morphine sulphate (Martindale Pharmaceuticals, UK) was repeated. Surgery ended without incident. Estimated blood loss was 1000ml. The epidural catheter was removed and the patient returned to ICU. Anticoagulant was to resume in 6hours.

CT scan of the chest and daily ABG were requested, but consent was declined. She was discharged to Neurologist for follow up on fifth day.

Discussion

The choice of anaesthesia for motor neuron disease is controversial partly due to rarity of cases to formulate guideline. Combined mini-dose spinal with sequential epidural anaesthesia in the sitting position was used successfully. Our patient's major challenge was severe dyspnoea and orthopnea. With patient unable to assume recumbent position, inducing general anaesthesia and inserting a supraglottic airway device as reported by Mistry et al. [6] was not feasible. As muscle relaxants especially depolarisers are contraindicated, instituting an IPPV via endotracheal intubation was not advised. Maroutti et al. [7] successfully used spinal anaesthesia in their patient. But our patient would not tolerate the sudden change in hemodynamic variables associated with spinal with her in the sitting position with the diaphragm splinted by uterus. We needed an anaesthesia that could allow the delivery of the baby and thus relieve the splint on the diaphragm while allowing improved ventilation. Sequential combined mini-dose spinal-epidural anaesthesia became handy. As patient's ventilation and saturation improved after delivery of the baby, the epidural catheter was loaded to extend anaesthesia. The resulting hypotension was successfully managed with ephedrine. Our method agrees with the work by Moreno-Gonzales in Colombia [8]. The morphine adjunct improved post-operative analgesia.

Due to no consent, radiological studies to diagnose specific respiratory and/or vertebral changes were not done.

Conclusion

In parturient with MND with severe dyspnea and orthopnea, subject to absence of contraindication, sequential combined mini-dose spinal-epidural anaesthesia was used with good outcome.

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