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Anaesthesia management in a dwarf patient with failed spinal anaesthesia and unanticipated difficult airway posted for emergency cesarean section: A case report

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Abstract
Background: Pregnancy in dwarfism generally present with contracted pelvis and cephalopelvic disproportion. Cesarean section is generally indicated for deliver. However, Anaesthesia management in dwarf pregnant patient is difficult and challenging as they are other concurrent conditions like atlanto-axial instability, spine deformities like kyphoscoliosis and narrowed pharynx leading to higher risk for both general as well as regional anaesthesia. Here, we are presenting a successful case of anaesthesia management in dwarf patient with failed spinal anaesthesia and unanticipated difficult airway for emergency cesarean section.

Keywords: Dwarfism, pregnancy, cesarean section

Introduction
The definition of dwarfism is considered to be height which is less than 2.3rd percentile of the CDC growth charts [1]. Dwarfism is defined as an adult height of less than 148 cm and is characterised by extreme and global growth failure· There are over 300 different types of genetic disorders of the skeleton classified currently, but these can be broadly divided into two groups: those with proportionate growth (short-trunk and short limbs) and those with disproportionate development (short limbs).Most women with skeletal dysplasia have a normal life-expectancy and fertility [2, 3]. The most common cause of dwarfism is a disorder called achondroplasia, which causes disproportionately short stature. Achondroplastic dwarfism in parturient may require cesarean section due to the accompanying cephalopelvic disproportion [4].

Case report
A thirty year old female with primipara with dwarfism, at 40 weeks of gestation with leaking amniotic fluid per vaginum presented for emergency caesarean section. On head to toe examination she had features of disproportionate dwarfism like a short torso and lumbar lordosis. She was 127 cm in height and weighed 45 kg. (BMI 27.9 kg/cu.m). She had protruding eyeballs and had frontal bossing and typical facies suggestive of achondroplasia. She gave positive family history of similar short stature in her elder sister. She was educated up to 10th standard and had normal intelligence. In airway examination she had 5 cm interincisor distance and was grade 4 via Mallampatti classification. Her thyro-mental distance was more than 6.5 cm. She had restriction of neck movements in the flexion-extension axis.

Her blood investigations were unremarkable. Her Electrocardiography (ECG) showed increased PR interval.

As the patient was being taken for an emergency cesarean section, further evaluation or work up was not possible and a decision to give regional anaesthesia with subarachnoid block (SAB) was taken. Written informed consent was taken for the same as well as the risks and complications associated with general anaesthesia explained and consent taken. The procedure explained to the patient. Patient was taken to operation room and standard ASA monitors (ECG, Non-invasive BP,Spo2,$ETCO_2$) applied and patients vitals noted heart rate(HR) 72 beats per min, respiratory rate(RR) 14 per min, BP was 130/70 mmHg, ECG

was showing increased PR interval,Spo2 showing 98% on room air. Wide bore 20 G intravenous (I.V) line secured. I.V ringer lactate (RL) started. The patient was pre-medicated with I.V Injection (inj) Pantoprazole 0.8 mg/kg; inj. Metoclopramide 0.2mg/kg, inj. ondansetron 0.08mg/kg induced with inj. Glycopyrrolate 0.004mg/kg. Under all aseptic precautions, SAB was given to the patient with 1.4 mL 0.5% hyperbaric inj. Bupivacaine with the addition of 10 mcg of Inj. Fentanyl as adjuvant. Drug was injected after confirming free, continuous and clear flow of cerebrospinal fluid and the negative aspiration of blood. Patient was immediately made supine and observed for 20 minutes. No motor or sensory block developed and no sympathetic response to spinal anaesthetic drug was observed. On further prodding, patient admitted to an alleged history of scorpion bite six years ago.

Fig 1: Body habitus of patient

Fig 2: Airway assessment

Considering past history and emergent situation, decision was taken to give general anaesthesia to the patient. Patient was preoxygenated with 100% oxygen. Inj. Propofol 2 mg/kg i.v. given. Once patient was under, Inj. Succinylcholine 2 mg /kg i.v. was given. Once fasciculations passed, rapid sequence (RSI) intubation attempt was made. However, the patient's jaw was not relaxed. Patient was ventilated with bag and mask ventilation for an additional two minutes and a bolus of 20 mg of Inj. Propofol was given and another attempt for intubation (RSI) was done resulting in the same event-unable to introduce scope in the oral cavity. As patient had starting desaturating to 90-93% of spo2, decision was taken to secure the airway with I-gel No.3 (second generation supraglottic airway device) as per the protocol of Difficult Airway Society. Air entry was checked and after confirmation by capnography, I-gel was secured and Ryle's tube introduced through gastric port. Anaesthesia was maintained with Oxygen (O_2) and Sevoflurane. Inj. Atracurium 0.75 mg /kg was used for neuromuscular blockade. Inj. Oxytocin drip 20 unit given after delivering baby. Inj. Midazolam 0.2 mg/kg and Inj. Fentanyl 2 mcg/kg were given and patient was maintenaned with O_2, Nitrous Oxide (N_2O) and Sevoflurane.

Once the surgery commenced, patient vitals were stable throughout the procedure. The surgery lasted for one hour 10 minutes. Once the procedure was completed, all anaesthetic agents stopped. Reversion of neuromuscular blockade was made with 0.05 mg/kg of inj. Neostigmine and 0.008 mg/kg of inj. Glycopyrrolate and patient extubated after regaining complete consciousness. She was monitored in the PACU for one hour and was observed to have zero spinal anaesthetic effect. The baby cried immediately after birth and had an APGAR score of 10 at 1 and 5 minutes respectively. No post-operative or post anaesthesia complications were observed.

Discussion

Achondroplastic dwarfism is the most common type of human dwarfism and is a genetic disorder of the growth plate of the bones. It has an incidence of 1 in 15000 to 1 in 40000 live births. Although it is an autosomal dominant genetic disorder, almost 80% cases occur with no family history. The characteristic findings of achondroplasia are the disproportionate short stature of limbs or trunk, exaggerated lumbar lordosis and typical facies associated with achondroplasia [5].

Hence, based on clinical findings, limited resources and emergent condition of the patient, a provisional diagnosis of achondroplasia was done and patient was managed accordingly although the exact cause for her short stature was not known.

The anaesthetic management of a patient of dwarfism for cesarean section is very challenging for anaesthesiologists as both regional as well as general anaesthesia pose risks in such patient. Anaesthetic challenges in patients with achondroplasia difficult neuraxial access with the unpredictable spread of local anaesthetic. The spine in patients with achondroplasia has several unique anatomical features, such as hypertrophy of superior and inferior articular facets, short and thickened pedicles of the vertebral body from premature fusion with scalloping on the posterior surface, prominent bulging of intervertebral discs, spinal canal tapering caudally from decreases in the interpedicular distance in the lumbar region (as opposed to widening caudally in normal anatomy), and thin dura. These mechanical features can lead to spinal stenosis and nerve root compromise. Determining the dose and volume of local anaesthetic is further complicated by accentuated lumbar

lordosis, spinal stenosis, engorged epidural veins, and narrow epidural and intrathecal spaces, which can result in the unpredictable spread or a high spinal/epidural block [6]. The pathophysiological diversifications of the different types of dwarfism as well as the difficult anatomical landmarks, spinal deformities and lack of proper guidelines for the management of patient with dwarfism for cesarean section are a few challenges faced by the anaesthesiologist [7]. In our patient, lumbar puncture was performed easily despite the anticipated difficulty. Due to her short stature, we carefully deliberated and decided to give her 1.4 mL of 0.5% hyperbaric inj. Bupivacaine with 10 mcg of inj. Fentanyl as adjuvant rather than the routine dose given for pregnant patients with average heights [8]. But due to non action of the spinal anaesthetic drug, we had to proceed with general anaesthesia. One possible reason for failure could be the lack of understanding regarding the subarachnoid volume or alleged history of bite by scorpion.

Another option is to perform general anaesthesia, in which case difficult airway places great risk due to the conditions such as big head and tongue, limited head extension, cervical spine instability and relatively undeveloped pharynx[9].Therefore, patients should be carefully evaluated to exclude difficult airway using indicators such as Mallampati classification, thyromental distance, sternomental distance. Different techniques for management of difficult airway should ideally be prepared in advance. For patients with high risk of difficult airway, awake intubation should be considered. Postoperative recovery may be affected when impaired cardiopulmonary function exists.

In the case of our patient, for general anaesthesia her restricted neck mobility and her reduced mouth opening and large tongue posed a challenge in securing a definitive airway [10]. Nevertheless, anaesthesia poses risks to patients with dwarfism, which should be recognised by all anaesthesiologists. Pre-operative assessments should be made carefully and contraindications such as severe spinal deformity and cord compression should be ruled out completely.

Conclusion

In our case report, we presented a case of successful anaesthesia management of a pregnant patient with dwarfism for emergency cesarean section. Patients with dwarfism pose significant challenge for both regional as well as general anaesthesia, especially in a low resource setting on an emergent basis. However, with careful consideration of available resources and various anaesthetic modalities, we successfully managed a pregnant patient with dwarfism and unanticipated difficult airway to have favourable maternal and fatal outcomes, thus providing insight into the management of such patients.

Declaration of patient consent

Written informed consent was obtained from the patient for publication of this Case report. A copy of the written consent is available for review by the Editor of this journal.

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Competing interests

The authors declare that they have no competing interests.

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