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Case Report

Anaesthetic management of an achondroplastic parturient for emergency LSCS

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Abstract
Achondroplasia, an autosomal dominant disorder, is one of the common causes of disproportionate dwarfism, with a worldwide prevalence of about 1:25,000-30,000. It is characterized by skeletal deformities, commonly affecting spine, secondary to genetic mutation in bone ossification. We present a case report of an achondroplastic primigravida with short stature, short neck, kyphoscoliosis and hypotonia of bilateral lower limb presenting for emergency LSCS. Anesthetic management of such cases can prove changing more so in emergency LSCS, owing to the physiological changes of pregnancy itself, which has been discussed in this case report.
Keywords: *Achondroplasia, Spinal anaesthesia, Caeserean.*

Introduction

Achondroplasia, an autosomal dominant disorder, is one of the commonest causes of disproportionate dwarfism; with a worldwide prevalence of around 1:25,000-30,000.[1,2] It is characterized by defect in bone ossification secondary to mutation in FGFR3 gene and its signaling causing numerous skeletal deformities including spine.[3] As such, management of a parturient for an emergency caesarean is challenging, owing to anticipated difficult airway and various physiological changes in different organ systems, additional deformities related to achondroplasia can further complicate the situation. We present successful management of one such case and the protocols that can help us deal with such difficult scenarios.

Case Report

A 29 -years old primi-gravida with short stature and kypho-scoliosis, at term-pregnancy presented for an emergency caesarean section in labour as normal vaginal delivery was not feasible due to significant cephalopelvic disproportion. She did not have any antenatal checkups done. Pre-anesthetic assessment was done. There was no history of short stature in her family or father of the child.

On examination, she was 110 cm tall and weighed 50 kg. She had a short neck with flexion of around 30-40^{0}and a negligible extension, kyphoscoliosis with fusion of lower lumber spine and hypotonia of bilateral lower limbs. Her mouth opening was 2 finger breadths with Modified Malampatti grade II. Her effort tolerance was around 4 METS with a breath holding time of 25 seconds. Her complete blood count was within normal limits. A written informed consent with a high risk for perioperative cardiopulmonary complications was obtained. She was premedicated with injection ranitidine 50 milligrams and injection metoclopramide 10 milligrams I.V for aspiration prophylaxis. We planned for a caesarean section under spinal anaesthesia. Difficult Airway cart and crash cart were kept ready.

For monitoring 5 lead ECG, non-invasive blood pressure and pulse oximetry were connected an 18 G I.V. cannula secured on dorsum of left hand with 20 ml/kg/hr Ringer lactate on flow. Under sterile aseptic precautions, spinal subarachnoid block was attempted in L_3- L_4 space in sitting position using a 25 G Quinke-Babcock needle. However, since the lower lumber vertebrae were fused and deviated from midline, we could not maneuver our needle through. Subsequently spinal anaesthesia was given in L_1-L_2 space. After obtaining free flow of CSF, injection 0.5% hyperbaric Bupivacaine 1.1 ml with 25 mcg injection fentanyl were administered. As the sensory level reached T_4, surgeons were asked to proceeded and a 2.5 Kg baby with apgar scores 8 and 9 at 1 and 5 minutes respectively was delivered. After 5 minutes her pulse rate dropped to 48 beats per minute and blood pressure was 90/ 50 mmHg. It was managed with 0.6 mg Injection Atropine I.V. bolus. Rest of the perioperative period was uneventful.

Figure 1 Lateral view of head and neck of the patient.

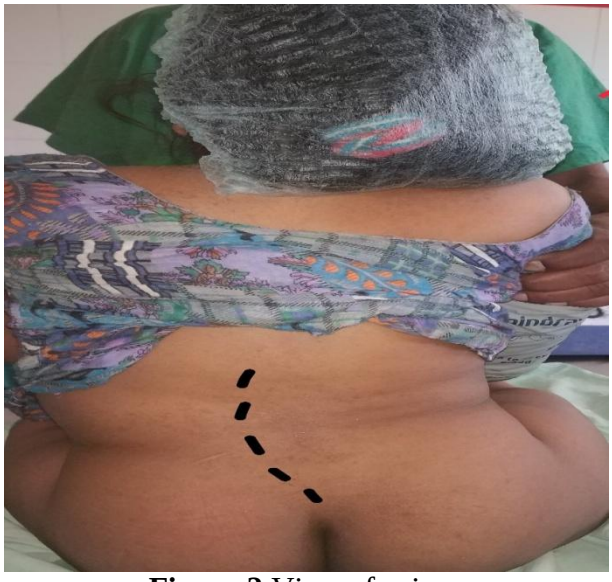

Figure 2 View of spine.

Discussion

Dwarfism, defined as height less than 147 cm in an adult, may be categorized as either **midgets** having trunk and limbs in normal anthropometric proportions or **dwarfs** having limbs longer than trunk or vice versa.

Achondroplasia is the commonest form of disproportionate dwarfism, with an autosomal dominant pattern of inheritance and female preponderance. Most of such cases result from a spontaneous mutation in FGFR3 gene leading to defective bone ossification.[3] These individuals usually present with abnormal maxillofacial morphology, spine deformities, elongated trunk

and shortened limbs.[2] They characteristically have low fertility rates and those who conceive and carry out their pregnancy till term; often present with challenges to anesthesiologist and obstetrician requiring caesarean section owing to CPD.[4] There is no single recommended anaesthetic approach for management of such cases. While in an emergency scenario, general anaesthesia is a commonly preferred technique; combined spinal- epidural may prove beneficial in elective scenario.[5,6,7] However, each modality has its own share of pros and cons.

As such pregnancy is an anticipated difficult airway owing to airway edema, achondroplastic individuals may additionally have abnormal maxillofacial and airway morphology including dental malocclusion, macroglossia, flat nasal bridge, difficulty in neck extension, atlanto-occipital instability; all of which lead to difficult bag and mask ventilation. Presence of macroglossia, nasopharyngeal stenosis, small trachea, narrowed rib cage and limited neck extension along with airway edema may also cause difficulty in or failure of endotracheal intubation. There is risk of spinal cord compression secondary to neck extension. Hence, a careful pre as well as postoperative neurological examination is deemed necessary in such cases.[8]

Perioperative cardiovascular complications may include pulmonary hypertension and perioperative MI. Pulmonary complications may occur in form of restrictive pulmonary pattern and obstructive sleep apnoea with reduced FRC which may deteriorate further, due to term gravid uterus pushing the diaphragm upwards. These patients may often have hypotonia of limbs due to spinal canal stenosis causing delayed recovery from muscle relaxant drugs. Thus, administration of general anaesthesia requires caution because recovery may be unpredictable. Moreover, pregnancy is considered a full stomach senario and if the patient is not adequately nil per oral as commonly seen in emergency caesareans, there is always a greater risk of aspiration.

Problems associated with epidural anaesthesia include difficulty in positioning and technique secondary to thoracolumbar kyphoscoliosis and lumbar hyper lordosis, increased chances of duralpuncture, bloody tap, patchy block, unpredictable sensory motor levels and even failed block. Similar are the complications associated with spinal anaesthesia in addition to dry tap, due to spinal canal stenosis.[9] Combined spinal-epidural anaesthesia may however be preferable over either of these techniques as we are not only able to administer titrated doses but also reduce the volume of spinal drug.

Mitra et al published a case report on anaesthetic management of patient with achondroplasia highlighting the benefit of using low dose opoid adjuvants to local anaesthetic solution, though not without taking proper precautions, as a tangible option for anaesthetic management of such cases.[10]

We planned our case under spinal anaesthesia even though the patient presented under emergency circumstances because of limitation of resources in our set-up to successfully counter the anticipated difficulties in general anaesthesia in such cases.

Learning Points

1) Anaesthetic management of achondroplastic parturients can prove challenging especially because there is no single preferred approach defined in literature for the same.
2) Each of the available anaesthetic techniques has its pros and cons.
3) A balanced anaesthetic approach with a defined algorithmic protocol must be devised keeping in view the skill and experience of anaesthesiologist and surgeon, patient profile and availability of resources in the said set-up.

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