Successful management of an Achondroplastic dwarf for Emergency Caesarean Section using Intrathecal Clonidine

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Abstract

Background: In day to day practice, on occasions we come across full term parturient, with concomitant genetic disorders. One such disease happens to be achondroplasia. We hereby discuss the successful anaesthetic management of a rare case of parturient with achondroplasia with difficult airway, who was scheduled for emergency caesarean section.

Methods: This being a clinical case report, the method of compiling and reporting was based on factual data of the cases in particular discussed with the further exploration of similar cases across the globe. Informed consent from the patient was taken, and subarachnoid block using clonidine as an adjuvant was given in lumbar space at the level L3-L4, with midline approach. A 2.0 kg male child was delivered with adequate APGAR score. Subsequently both mother and child were shifted to PACU.

Result: The general recommendation regarding the ideal anaesthetic technique can't be given as both general and regional anaesthesia present problems, thus individualized decision is necessary. In our case, spinal anaesthesia was planned, however it was technically difficult, since her spine was kyphoscoliotic and risk of patchy block was there, but, since the patient was full stomach, we preferred it. Here, we used clonidine as an adjuvant with low dose of hyperbaric 0.5% bupivacaine to achieve adequate level of block. It significantly prolongs postoperative pain relief and there is no impairment of APGAR score of the baby with this drug.

Conclusion: The risk of general anaesthesia and regional anaesthesia in achondroplastic patients are known, so complete evaluation before administration of anaesthesia is needed to reduce risks. The plan of anaesthesia should be individualised after detailed risk benefit analysis. With meticulous planning Subarachnoid block can be successfully administered to a parturient with Achondroplasia for caesarean section.

Keywords: Achondroplasia, primigravida, spinal anaesthesia.

Introduction

There are several genetic disorders in pregnant females which can create challenges to anaesthesiologists and obstetricians. Achondroplastic dwarfism is one of these disorders whose management always remains controversial. It is an autosomal disorder and is a commonest form of rhizometric dwarfism [1] with abnormal endochondral ossification. It is associated with several anatomical changes and physiological changes [2], making the anaesthetic management of such a patient a challenge more so when a paturient with achondroplasia presents for a caesarean section. Administration f either general or regional anaesthesia is associated with its own share of complications, making it a tough choice for an anaesthesiologist[3-6] .We hereby discuss the anaesthetic management of a rare case of parturient with achondroplasia with Mallampatti grade 3, mouth opening only for two fingers who was scheduled for emergency caesarean section. The patient was successfully managed by administering spinal anaesthesia using intrathecal clonidine as an adjuvant with low dose hyperbaric bupivacaine, and thus highlighting its safety and efficacy for such patients.

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Case report

A 19 years primigravida, clinically diagnosed as a case of achondroplastic dwarf with 38 weeks of pregnancy, was posted for an emergency Caesarean section for cephalopelvic disproportion with leaking per vaginum. She had taken her meal one hour before coming to the hospital, otherwise her previous medical history was not significant. On general examination the patient was short stature, with small limbs; her height was 105 cm and weight being 40 kg. On airway examination she had Mallampatti grade 3 and mouth opening was two fingers, with retracted mandible. Hence it was an anticipated difficult airway. On examination of spine it was kyphoscoliotic at the thoracolumbar level, however the lower lumbar spaces were moderately palpable.. Her cardiovascular and respiratory system examination were unremarkable. Her baseline hemogram was 11g/dl and other investigations could not be done due to paucity of time. The patient was premedicated with ranitidine 50 mg IV and metoclopramide 10 mg IV preoperatively; meanwhile preparations for the anticipated difficult airway were made in the operating room. The patient was shifted in the OR, connected to multipara monitor and was monitored for SpO2, non invasive blood pressure, heart rate. The patient had taken meals 1 hr before, considering high risk of aspiration and suspecting difficult intubation, spinal anaesthesia was planned for caeserean section and injection of mephentermine as well as injection of atropine were kept ready to manage any event of hypotension and bradycardia. The minimum values of MAP(mean arterial blood pressure) and HR(heart rate) that were acceptable in our case were 60 mm Hg and 50/ min respectively.

Informed consent from the patient was taken, and she was preloaded with 500 ml Ringer lactate through a 18 G cannula prior to procedure. The patient was placed in sitting position and subarachnoid block (SAB) was given in a single attempt with 26 G Quincke needle by midline approach at L3-L4 interspace after obtaining free flow of cerebrospinal fluid. Keeping in mind the short stature (105cm) of the patient, a total of 7.5 mg of 0 .5 % hyperbaric bupivacaine with 20μg clonidine as an adjuvant was used to produce block. The patient was made supine with a fifteen degree left lateral tilt, and pillow of height 10 cm was kept under head to restrict height of block. Sensory analgesia up to T4 -T6 dermatome was confirmed and her vitals were recorded initially every 2 min for 15 min and then recorded every 5 min till the completion of surgery .The total blood loss was 400 ml approximately and total of 1.5 litres of fluid in the form of Ringer lactate was given throughout the surgery. The procedure lasted for 70 min, where the patient had stable hemodynamics and oxygen saturation. A 2.0 kg male child was delivered with APGAR score of 7and 9 at 1 and 5 min. Subsequently both mother and child were shifted to PACU. In the postoperative period analgesia lasted for 7.5 hrs and the patient remained hemodynamically stable. Thereafter intravenous paracetamol 1 gm infusion TDS was advised for pain relief and the remaining course of the patient in the hospital was uneventful. Finally the patient was discharged from the hospital 6 days later following procedure.

Discussion

Achondroplasia is the most frequent of more than 100 described type of skeletal dysplasia which leads to dwarfism [2]. These patients have a number of anatomical and physiological abnormalities that contributes to problems with anaesthesia in these patients. Achondroplasia, genetically is due to mutation of the FGF receptor gene [7]. This mutation results in an inhibition of cartilage proliferation and disorder of enchondral ossification, as a consequence premature ossification is observed. Clinically, the following symptoms are characterstic: disproportionate dwarfism, a relatively large head, midfacial hypoplasia, deformation of the spine. As primary or secondary consequences, other organ system can be affected [2].

A general recommendation regarding the ideal anaesthetic technique can't be given as both general and regional anaesthesia present problems thus individualized decision is necessary. General anaesthesia poses significant challenges such as, excessive anxiety [7] and increase risk of cardiovascular complications [8]. These patients also pose with difficult airway, listing some of these like: difficult intubation [9, 10, 11], pharyngeal and maxillary hypoplasia, associated with large tongue, narrow nasal passages, chronic respiratory infection [2] and restrictive lung disease. The base of skull is shortened and angulated, yielding difficult intubation. Increase tendency of sleep apnea is there due to craniofacial abnormalities and hypotonia of upper airways muscles [12]. In our case also, since patient had taken meals and had anticipated difficult airway, we preferred SAB. As the patient was in active labor and leaking P/V, and SAB being the commonest technique with a rapid onset and a definite end point, it was preferred. Epidural anaesthesia or CSE were not preferred, because it is technically difficult in these cases [6]. Accidental dural puncture is quite common here and an inadvertent intrathecal injection could lead to high blockade [1].

However in our case, spinal anaesthesia was also technically difficult, since her spine was kyphoscoliotic and risk of patchy block was there, but it was felt that since the patient was full stomach so it would be a better option than general anaesthesia. In this patient, we used 20 μg clonidine as an adjuvant along with low dose of 5% hyperbaric bupivacaine to achieve adequate level of block. It has been studied that clonidine is a safe choice as an adjuvant, intrathecally in cesarean section [13], it provides effective analgesia and motor paralysis at a lower dose of bupivacaine. It also significantly prolongs postoperative pain relief and there is no impairment of APGAR score of the baby with this drug [13]. However appropriate precautions were taken to restrict height of block and every preparation of emergency tracheal intubation in this patient if any complication arises, were kept.

It has been studied and reported in the past, that regional anaesthesia is also technically difficult, due to anatomical changes in the spine (narrow spinal canal, stenosis, reduced epidural space, kyphoscoliosis, and vertebral body deformities [14]). Recognition of SAB tap may be problematic, because free flow of CSF may be difficult to obtain [14], and the risk of high spinal anaesthesia is also conceivable [15]. . In one case report, Derenzo et al [5] reported failed regional anaesthesia with reduced dosage of spinal bupivacaine in achondroplastic parturient. Even the insertion of catheter is difficult because of presence of narrow epidural space [6]. Engorged epidural veins, increases the risk of venous puncture either by the Tuohy needle or the catheter, and results in an segmental block spread of local anaesthetic within the space [11]

Generally speaking, general anaesthesia has been the preferred technique for caesarean section in achondroplasia, however a few case reports of successful management by regional anaesthesia technique have been published in literature, including spinal[1,5,6] , epidural[7,11] or CSE [10] . Here in our case report, an achondroplastic patient presented for an emergency cesserean section, where we used clonidine as an adjuvant for the first time. In one of these reports on regional anaesthesia, as above mentioned, Sukanya et al [1] reported successful use of spinal anaesthesia in emergency caesarean section of achondroplastic female, where she used fentanyl as an adjuvant along with hyperbaric 0.5% bupivacaine. In another case report, Trikha et al [10] preferred CSE over a spinal or epidural tecnique because it combines the rapid onset of spinal aneasthesia with the implantation of catheter. Wardell and Frame et al [11] reported that only 5 ml of 0.5% plain bupivacaine in epidural was sufficient to achieve block up to T4.

There always been a dilemma for the anaesthesiologist, to select type of anaesthesia, as decision may be guided by various factors such as anticipated difficult airway, indication of caesarean section emergency or elective, previous history of any associated comorbidities, any organ involvement or atlantoaxial instability to avoid general anaesthesia. In this case report, we successfully managed the caesarean section under spinal anaesthesia, using clonidine as an adjuvant with stable hemodynamics intraoperatively as well as in postoperative period. The management also highlights the analgesic safety and efficacy of clonidine as an adjuvant with duration of analgesia lasting for 7.5 hrs.

Conclusion

Although subarachnoid block is not being commonly practiced by several practitioners in Achondroplastic patients, even there are no prescribed standard dosage guidelines for spinal anaesthesia in such cases; hence it should be used with caution. However the chances of successful spinal anaesthesia can be increased by careful selection of the patient and appropriate preparations accordingly.

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