Emergency Cesarean Section in a Patient with Achondroplasia: An Anesthetic Diliemma

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ABSTRACT

Background: **Achondroplasia, the commonest form of short-limb dwarfism, is associated with several bony changes in face, neck, spine, and can also have neurological and cardiopulmonary complications. Thus it presents several challenges to the anesthesiologist as to the best course of action, and there is an ongoing debate regarding general vs. regional anesthesia in these patients. In particular, there are very few reported cases of spinal anesthesia due to its feared high risks.**

Case report: **We report a case where spinal anesthesia was used for emergency Caesarean section in an achondroplastic woman, 109 cm tall and weighing 45 kg, with 37-week pregnancy presenting with cephalopelvic disproportion and fetal distress. She had mild lumbar lordosis but no other spinal or systemic abnormality. Low-dose bupivacaine and fentanyl were used. There was no technical difficulty encountered while doing the procedure, and an adequate block (but not too high) was achieved.**

Conclusion: **We discuss anesthetic issues and provide some general guidelines in dealing with achondroplastic patients. The role of regional anesthesia, especially spinal anesthesia and a dosage guideline is highlighted. Under favorable circumstances and if urgently needed, spinal anesthesia remains a viable option in these patients.**

KEY WORDS: Achondroplasia; Dwarfism; Anesthesia: Regional; Anesthesia: General; Anesthesia: Spinal.

Short-limbed dwarfism involves shortening of distal, middle or proximal part of the limbs (acromelic, mesomelic, and rhizomelic subtypes, respectively). Achondroplasia is the commonest form of rhizomelic dwarfism. This autosomal dominant disorder is associated with abnormal endochondral ossification whereas periosteal and intramembranous ossifications are normal. These patients have peculiar facial features, bony deformities and systemic abnormalities that often make administration of anesthesia challenging.[1] There are several uncertainties regarding the mode of anesthesia, exact procedures and protocols, drug choice and dosage, etc.[2-5] There is only one published report from India.[6] Amongst regional anesthetic techniques, in contrast to epidural anesthesia where a number of published reports are available, reports of successful spinal anesthesia in achondroplastic dwarfs are controversial and rare.[7,8]

Here we report a case in which we successfully used spinal anesthesia with low dose bupivacaine and fentanyl combination in a woman with achondroplasia undergoing emergency Caesarean section, discuss the related issues, & provide some general guidelines for the anesthesiologist, especially highlighting the role of regional, and, in particular, spinal, anesthesia for these patients.

CASE REPORT

A 25-year-old nulliparous achondroplastic dwarf with 37-weeks of pregnancy was admitted for an emergency Caesarean section for cephalopelvic disproportion with fetal distress. She was otherwise normal and her previous medical history was unremarkable. On examination, her height was 109 cm and weight 45 kg. She did not have any obvious spinal deformity other than mild lumbar lordosis. Cardiovascular

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and respiratory system examination were unremarkable. Her airway examination was unremarkable other than the presence of protruding upper incisors. She was placed in Mallampati grade 2. Her preoperative baseline investigations were within normal limits. and she received premedication of ranitidine 50 mg iv and metoclopramide 10 mg iv preoperatively.

In the operating room she was connected to multichannel monitor (Datex Light, Helsinki) and was monitored for pulse oximetry (SpO2), noninvasive blood pressure (NIBP), electrocardiogram (ECG) and end tidal carbon dioxide (EtCO2). She was also preloaded with 500 ml normal saline prior to the procedure. The patient was placed in the left lateral position and subarachnoid block was given with 26 G Quincke needle at L3-L4 interspace after obtaining free flow of cerebrospinal fluid. 1 ml of 0.5% bupivacaine (heavy) with 10 µg Fentanyl (0.2 ml) was used to produce the block. The patient was made supine with a fifteen degree left lateral tilt. Sensory analgesia up to T4-T6 dermatome was confirmed and she remained pain-free throughout.

A 2.4 kg achondroplastic female child was delivered with Apgar scores of 7 and 9 at one and five minutes respectively. Her vitals remained stable throughout the course of surgery. The surgery lasted for 1h and she received 1.5L of normal saline during this period. Her estimated blood loss was 250 ml and she required no vasopressor during the entire course of surgery. At the end of the procedure the mother and child were shifted to the post anesthesia care unit (PACU) where they had an uneventful postoperative period. They were discharged from the hospital 5 days later.

DISCUSSION

Various factors have been noted to be of importance in deciding the best course of action regarding choice of anesthetic technique in these patients (Table). A proper preanesthetic evaluation can prepare an anesthesiologist for anticipating potential complications.

Both general as well as regional anesthesia in these patients are fraught with problems. One needs to be well aware of this (Table) before taking up these patients for surgery and anesthesia. There is a notion that general anesthesia is the preferred technique.[1,2] It is not clear, though, if this recommendation is based on a documented superiority of general anesthesia or rather due to an unease and reluctance to perform regional anesthesia on these patients. Epidural anesthesia has been reported to be successful in a number of reported cases or case series.[6, 9-15] Although successful, there are earlier reports of technical difficulty and even incidence of dural puncture.[1, 2, 10] A high level of block is another complication reported with this technique.[11] Further, dosage guidelines are unclear due to non-uniformity of the spinal canal lumen and unpredictability of spread of the drug through the epidural space; thus, both the type and volumes of the ideal drug remain unclear.[15]

Spinal anesthesia, on the other hand, has rarely been used, with only 4 published case reports till date.[5, 7,8,16] Of these, Crawford and Dutton[7] used a 32-gauge microspinal catheter for continuous spinal anesthesia, but these catheters have later been withdrawn due to concerns regarding causing neurological deficits.[15] Trikha at al.[16] used combined spinal-epidural anesthesia rather than only spinal. A lack of proper guidance regarding dosage of drug to be used with spinal anesthesia was one reason for their choice of this technique. De Renzo et al.[5] reported need for supplementary intravenous sedation after inadequate spinal anesthesia in an achondroplastic patient with complex needs.

One website-based medical guideline (published in the website for the "Little people of Ontario") says, "Specifically, spinal anesthesia should not be used in achondroplasia", and, instead, recommends use of general anesthesia for achondroplastic patients undergoing caesarian section.[17] An uncertainty over spread of local anesthetic drug through the stenosed spinal canal and raising the possibility of a high spinal block may have caused reluctance to recommend spinal anesthesia in these patients. However, other than hypotension, no other serious adverse events, including the much-feared neurological deficits, have so far been reported.[15]

` It may be worth mentioning that bony changes in patients with achondroplasia are similar to those in elderly and spinal anesthesia with low-dose fentanyl and bupivacaine often remains the technique of choice in this subset of patients. Monedero et al.,[4] in their review of a case series, commented that fears

Table

Characteristics of achondroplastic patients that can influence the anesthetic choice, procedures and complications. [Note that these features are not necessarily present in each case, and when present, can vary widely in severity.] GA: general anesthesia; EA: epidural anesthesia; SA: spinal anesthesia.

System	Characteristics	Issues for GA	Issues for EA	Issues for SA
Bones	Maxillary hypoplasia, large mandible, megalocephaly with protuberant	Difficulty in mouth opening and visualization of glottis		
Face, Neck, Skull	forehead, flat nose, macroglossia, difficulty in exposing the glottis, narrow nasal passages & nasopharynx	due to macroglossia Endotracheal tube size		
Short neck	Limited neck extension	and risks of forced		
	Marked cervical kyphosis	extension		
	Fusion of the atlanto-occiptal articulation	Difficulty in sealing facial mask due to peculiar facial features		
	Foramen magnum stenosis			
Bones	Thoracolumbar kyphoscoliosis		Difficulty in positioning	As for EA
Spine	Relatively narrow spinal canal		Difficulty in finding landmarks	Difficulty in obtaining free flow of CSF due to spinal canal stenosis
	Vertebral deformities (shortening of pedicles, decreased interpedicular distance especially in lower lumbar spine, osteophyte formation)		Difficulty in catheter/ needle insertion	
Bones	Rib hypoplasia, with flattened rib cage	Difficulty of midline positioning of laryngoscope due to pectus carinatum		
Others	Pectus carinatum or excavatus			
	Genu varum			
Neurologic	Foramen magnum stenosis	Risk of cervico-medullary compression		
	Sleep apnoea	Risk of hyperthermia		
	Hydrocephalus, with increased intracranial pressure			
Cardiopulmonary	Restrictive lung disease	Possible complications due to preexisting lung and/or heart disease		
	Pulmonary hypertension Cor pulmonale (from pulmonary hypertension, restrictive lung disease and apnea)			
Miscellaneous		Unclear dose requirements for anesthetic and muscle relaxant	Dosage guidelines unclear	Risks of inadvertent high spinal block , e.g., profound hypotension
Decreased height & weight				
Disproportionate dwarfism				
Non-uniform spinal canal lumen				

associated with anesthesia in achondroplasia might have been excessive; this might be applicable in the case of spinal anesthesia as well.

In our case, the choice of spinal anesthesia was dictated by several factors. First, general anesthesia in this emergency surgery was deemed to be risky, especially due to lack of adequate preparation of the patient, risk of aspiration, along with the usual risks for general anesthesia in a patient with both achondroplasia and advanced pregnancy. Further, other than a mild lumbar lordosis, the patient did not have any other spinal deformity and hence was not unsuitable for regional anesthesia. We decided for spinal anesthesia rather than epidural due to the urgency demanded by the situation.[5,8] As reported earlier, a low-dose combination of bupivacaine and fentanyl was sufficient and safe.[8] We, however, used a lower dose of bupivacaine with fentanyl in our patient and level of analgesia was adequate with this dose regime. Further our patient did not require any vasopressor during the course of surgery. Hence 1ml of bupivacaine with 0.2 ml of fentanyl seems safe and adequate in achondroplastic pregnant patients for spinal anesthesia.

Thus, our case report has an educational value in documenting the fact that spinal anesthesia with low-dose fentanyl and bupivacaine, administered carefully while being aware of potential complications and with due precautions, can also be a tangible option in anesthesia for achondroplastic patients. A precaution is not the same as a contraindication and least so in patients where general anesthesia is not safe or feasible. We feel that spinal anesthesia thus remains a viable option in management of achondroplastic patients undergoing emergency caesarian section.

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