Combined spinal epidural anesthesia in achondroplastic dwarf for femur surgery

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Abstract

Achondroplasia is the commonest form of short-limbed dwarfism and occurs in 1:26,000-40,000 live births. This is an autosomal dominant disorder with abnormal endochondral ossification whereas periosteal and intramembranous ossification are normal. The basic abnormality is a disturbance of cartilage formation mainly at the epiphyseal growth plates and at the base of the skull. The anesthetic management of achondroplastic dwarfs is a challenge to the anesthesiologist. Both regional as well as general anesthesia have their individual risks and consequences. We report a case of an achondroplastic dwarf in whom combined spinal epidural anesthesia was used for fixation of a fractured femur. The patient had undergone previous femur surgery under general anesthesia since he had been informed that spinal anesthesia could be very problematic. There was no technical difficulty encountered during the procedure and an adequate level was achieved with low-dose local anesthetics without any problem. Postoperative pain relief was offered for three consecutive postoperative days using epidural tramadol. We discuss the anesthetic issues and highlight the role of combined spinal epidural anesthesia with low-dose local anesthetics in this patient. This approach also helped in early ambulation and postoperative pain relief.

Introduction

Dwarfism is defined as failure to achieve a height of 148 cm by adulthood.[1,2] People with severe short stature have been classified as those with normal ratio of trunk to limb length (proportionate dwarfism) and those with disproportionate development. Achondroplasia, the commonest form of short limb dwarfism, is associated with several bony changes in the face, neck and spine, and can have neurological and cardiopulmonary complications. An exaggerated lumbar lordosis, thoracic kyphoscoliosis, generalized spinal stenosis and the unpredictable behavior of local anesthetic drugs in epidural and spinal space have led to a reluctance among anesthetists to consider regional anesthesia in this group of patients.[3] Craniofacial abnormalities, a large head, saddle nose, limited neck extension, foramen magnum stenosis, a large tongue and mandible, and atlanto-axial instability can make general anesthesia potentially hazardous. Thus it poses several challenges to the anesthesiologist as to which is the best course of action.

Case Report

A 56-year old male patient, height 134 cm, weight 45 kg, was programmed for fixation of right fracture neck femur. He had an arm span of 127 cm, upper segment of 78 cm and lower segment of 56 cm confirming disproportionate dwarfism. His mother was an achondroplastic dwarf. He had a past history of left side fracture femur for which plating was performed under general anesthesia 12 years ago. At that time, the patient was informed that spinal anesthesia could be problematic, so surgery was performed under general anesthesia, which was uneventful. But the patient experienced a lot of postoperative pain, nausea and vomiting. The patient had a large head, short limbs, a saddle nose, bilateral bowing of legs and mild kyphoscoliosis of the thoracolumbar spine. Lumbar intervertebral spaces were well palpable. Mallampati classification was grade II and neck movements were normal. All preoperative investigations were within normal limits. Considering the nature, duration and site of surgery, and the potential risks and benefits of general anesthesia compared with regional anesthesia, it was decided to perform the operation under combined spinal epidural block.

On the day of surgery, routine monitors were attached. These included: cardioscope, pulse oximetry, non-invasive blood pressure monitor and urine output. An 18-gauge IV cannula was placed and Ringer lactate was started. A 16 gauge Epidural catheter was placed in the L2-L3 space using loss of resistance technique. Subarachnoid block was given with a 25 gauge spinal needle in theL3-L4 space. Then 1.5 mL (7.5 mg) of heavy 0.5% bupivacaine was injected. The height of block achieved was T8. Vital parameters were maintained within normal limits. At 90 min., the level receded to T10 and 2 mL of 2% lignocaine adrenaline (1:2,00,000) was given as test dose. At 120 min., 3 mL of 0.5% bupivacaine was given. Thereafter no top up was required until the end of surgery which lasted for 3 h. Postoperative epidural analgesia was achieved with an injection of tramadol 50 mg diluted in 10 mL 0.9% normal saline, as required for three consecutive postoperative days. The postoperative course was uneventful and the patient was discharged on Day 10 after surgery.

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Discussion

Achondroplasia, the commonest form of short-limb dwarfism, is associated with several bony changes in face, neck and 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 is an ongoing debate regarding general *versus* regional anesthesia in these patients. Anesthesia, whether general or regional, presents many potential problems in achondroplastic dwarfs. Maintanence of an upper airway may be difficult because of the presence of a large tongue and a large mandible. Tracheal intubation may prove difficult as foramen magnum stenosis and cervical spine instability are common and hyperextension of the neck must be avoided to prevent cervical cord compression. Perovic *et al.* found that 75% of non-achondroplastic dwarfs have atlanto-axial instability and hence care must be taken during intubation.[4] Limited neck extension has been reported as the cause of difficult intubation in 2 cases[5,6] although not all authors have reported similar difficulties. Mayhew and colleagues[7] reported no difficulty in airway management or direct laryngoscopy in a series of 27 patients undergoing 36 anesthetic procedures. If tracheal intubation is required in this group, a small tracheal tube should be selected, the most appropriate size being estimated according to weight rather than age. Our patient underwent his previous femur surgery under general anesthesia as he had been informed that spinal anesthesia could pose problems. Following GA, the patient experienced severe pain, nausea and vomiting during that postoperative period. Since the patient had a mild thoracic kyphoscoliosis with

a normal lumbar spine we advised regional anesthesia. Weighing the risks and benefits of both regional and general anesthesia, the patient expressed his desire to be awake and agreed for regional anesthesia, especially after his previous unpleasant experience of general anesthesia. Regional anaesthesia may be technically difficult because of poor landmarks, but many achondroplastic dwarfs have a normal spine. A relatively narrow spinal canal may result in cord compression, prolapsed discs and deformed vertebral bodies.[8,9] In elderly achondroplastics, the possibility of disc herniation with resultant paraplegia may warn against the use of subarachnoid blockade.[10] A narrow epidural space may make catheter insertion difficult and a subarachnoid tap more likely. Recognition of subarachnoid tap may be problematic because free flow of CSF may be difficult to obtain.[8] Although the spinal abnormalities in dwarfs may increase the technical difficulties of administering regional anaesthesia, one should note that these abnormalities are similar to the degenerative changes commonly seen in the elderly, a population in whom regional techniques are frequently used.

Amongst regional anesthetic techniques, in contrast to epidural anesthesia for which a number of published reports are available, reports of successful spinal anesthesia in achondroplastic dwarfs are controversial and rare.[11,12] A Combined Spinal Epidural (CSE) technique was preferred over spinal or epidural anaesthesia because it offers rapid onset of dense anesthesia of subarachnoid block and also the flexibility of extending anesthesia through the epidural with titrated doses of drugs and postoperative analgesia. We used low-dose of bupivacaine (7.5 mg) to reduce complications and neuraxial block was extended with small incremental doses of epidural bupivacaine. There are reports of low epidural dose requirements in dwarfs. The most appropriate type and volume of epidural test dose is unclear. Plain 0.5% bupivacaine in doses of 2 and 3 mL have been reported.[13] Each top up should be used as test dose to avoid high-level blockade in these groups of patients. Brimacombe and Caunt reported the development of an epidural block extending from C5 to S4 over a 20 min. period using 0.5% plain bupivacaine 12 mL,[14] while Wardall and Frame reported that only 5 mL of 0.5% plain bupivacaine was sufficient to develop a block to T4.[13]

It is routine practice in our institute to give spinal and epidural with a two level approach in which the epidural cathetor is placed one space above the spinal block. In terms of cost, this is more economical to most of our patients who are from lower socioeconomic groups as combined CSE sets are very expensive. The concerned patient in our case report was poor, and since lumbar spaces were well palpable as already mentioned we proceeded with the routine two level approach. Moreover, studies comparing the needle through needle technique with separate needle technique have found a higher rate of failure of the spinal component with the needle through needle technique.[15,16] Failure rates of 5-20% have been reported for the needle through needle technique compared with less than 5% for the separate needle technique. There have been few studies on the rates of failure of the epidural component. Occasio-nally problems may be encountered with inserting an epidural catheter following the spinal approach, resulting in a significant delay between the spinal and epidural components of the CSE. Such delays may result in the spinal component of the block becoming fixed before the anesthetist has had a chance to position the patient. Risk of the epidural catheter being accidently inserted into the subarachnoid space via the hole created by the spinal needle is a rare complication but this risk is greatest with a needle through needle technique. This technique does not result in shearing of epidural catheter as literature shows that catheter shearing is generally seen due to excess force while removing the catheter or withdrawing the catheter back through the needle or withdrawing it over a deformed or damaged needle bevel.[17] It has been suggested that regional anesthesia should be avoided in patients with achondroplasia because any subsequent neurological abnormalities caused by spinal deformities may be attributed to the anesthetic technique.[5] But no neurological abnormalities in patients with achondroplasia have yet been reported in those cases performed under regional anesthesia. As there were no pre-existing problems in our patient, it was felt that there were no specific contraindications to the use of spinal and epidural anesthesia. In common with previous experiences, no neurological sequelae developed in our case. In summary, the characteristics of achondroplastic patients can influence the anesthetist's choice, all features are not necessarily present in each case and can vary widely in severity. The plan for general or regional anesthesia should be based on each individual case. Combined spinal epidural anesthesia is a viable option in the management of achondroplastic patients. Low dose of local anesthetic with an epidural catheter *in situ* can successfully be used in such patients without adverse effects and postoperative pain relief can be offered which is beneficial for early ambulation of elderly patients.

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