

USG Guided Combined Spinal Epidural: A Boon in an Achondroplastic Dwarf undergoing an Orthopaedic Procedure

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Abstract

Achondroplasia is one of the commonest forms of dwarfism. A 26-year-old achondroplastic dwarf with vitamin D deficiency was posted for an elective shaft of femur fixation. Owing to her difficult airway and past history of tuberculosis, it was decided upon to operate her under combined spinal and epidural anaesthesia. This case report highlights on the need for real time ultrasound guidance in locating the epidural space and hence aiding in successful placement of the Quincke's needle and the epidural catheter. The ultrasound is an effective and reliable tool to ascertain the most feasible inter-vertebral space in cases of a difficult spine anatomy.

Keywords: Achondroplasia; Ultrasound; Difficult spinal

Introduction

Achondroplasia is an autosomal dominant disorder caused by sporadic mutation of the *FGFR 3* gene leading to abnormal enchondral ossification. It is characterized by disproportionate dwarfism, a relatively large head, midfacial hypoplasia, deformations of the spine, leg axis deviation, spinal canal stenosis, kyphoscoliosis, reduced epidural space and vertebral body deformities posing a challenge to both region and general anaesthesia [1]. Anaesthetic management of achondroplastic patients undergoing a variety of elective and emergency surgical procedures have been reported, the most common include limb lengthening surgeries [2], Caesarean section [3], bariatric surgery [4], laparotomy for bilateral oophorectomy [5], vascular repairs [6] and middle ear surgeries [7]. Neuraxial block can be a challenge and hence the use of real time ultrasound could mitigate unsuccessful epidural insertions.

Case Report

A 26-year-old female presented with a fracture of the right femur for which a femur fixation was planned. She was a known case of achondroplasia and vitamin D deficiency, chronically immobile with a history of fall leading to pain and swelling of her hip. She was an old case of pulmonary tuberculosis for which she was adequately treated. On physical examination she was measured to be 118 cm with a weight of 40 kg. She had a large head disproportionate to her body with short neck, mandibular protrusion and a kyphoscoliotic spine. Airway examination showed an MPC III, restricted neck movements, a thyromental distance of 5 cm and adequate mouth opening. Cardiovascular system examination was normal with a heart rate of 68/min and a blood pressure of 110/70 mm of Hg. Respiratory system examination revealed a depressed chest wall with a protuberant abdomen and thoracic kyphosis, auscultation of the chest was unremarkable. An endocrine opinion was taken in view of vitamin D deficiency (\downarrow vitamin D, \uparrow PTH) and optimization of vitamin D and calcium were done prior surgery. Cardiac and pulmonary medicine work up was also done. Her biochemical profile and blood profile were within permissible limits with the exception of her hemoglobin being 9.3 g%. Electrocardiogram showed bilateral ventricular hypertrophy which was ensued by 2D echo wherein the ejection fraction was 60%. Arterial blood gas analysis showed a PH -7.42, PCO_2 -37, PaO_2 -108 and saturation of 98% on room air. Central neuraxial block was planned on this patient in view of anticipated difficult intubation and previous history of pulmonary tuberculosis. The patient was explained the procedure of a combined spinal epidural and the chances of a multiple/failed attempts and the need for conversion to general anaesthesia. Standard anaesthesia care of electrocardiogram, pulse oximetry, $ETCO_2$

and noninvasive blood pressure were used to monitor the patient. Intravenous access was obtained on the right upper limb using a 20G IV cannula. The patient was preloaded with 250 mL of ringer's lactate. The patient was given a left lateral position. Ultrasound was used to identify the intervertebral space and hence the spaces for epidural catheter placement and subarachnoid block. The best space for epidural and the distance to reach the epidural space was calculated.

Epidural space was obtained at L1 L2 intervertebral space and the catheter was fixed at 6 cm. Spinal was given at the L3 L4 intervertebral space. 2 cc of 0.5% bupivacaine heavy was injected using a 25 g Quincke's needle. Post subarachnoid block the patient was given head high to prevent high spinal levels. Despite head high a pre-operative spinal level of T6 was obtained. The procedure lasted for 3½ hours. Epidural was activated using 2 mL of 2% preservative free lignocaine and thereafter an infusion of 3 cc of 0.375% bupivacaine was started. Patient was hemodynamically stable throughout the procedure. Blood loss of around 350 mL occurred during the procedure and was replaced. Post procedure spinal level was L1 and the patient was comfortable and pain free. The epidural catheter was removed post operatively as the aim of our epidural was only to prolong the duration of anaesthesia (Figure 1).

Discussion

Achondroplasia also known as short limb dwarfism has an incidence of 1 to 1.5 /10000 live births [8]. It is inherited in an autosomal dominant fashion with the incidence being higher amongst females. Spontaneous mutation of the *FGFR 3* gene leads to enchondral ossification of the epiphyseal growth plate [8,9]. An achondroplastic patient can pose significant challenges to the anaesthesiologist. Hence preoperative assessment and planning of anaesthesia are of foremost concerns. Routine biochemical profile, lung functions, echocardiograph and ABG analysis have to be sought for in relevant cases [10]. It is prudent to ask for a neurological consult in cases with cervicomedullary stenosis and

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Figure 1: Ultrasound image.

cervical canal stenosis. General anaesthesia can be a challenge to the anesthetist owing to difficult mask ventilation, difficult intubation with regards to relatively larger heads, midfacial hypoplasia, restricted neck movements, cervicomedullary stenosis and cervical canal stenosis [11]. Regional anaesthesia in dwarfs could be complicated by the presence of ill-defined anatomical landmarks, spinal stenosis, osteophytes, short pedicles or a small epidural space. This could lead to multiple attempts at getting to the epidural space, increased risk of dura puncture, erratic spread of the local anesthetic and patchy action owing to the smaller space and stenosed canal. A number of authors and studies have warranted the use of general anesthesia owing to its safety profile, but we opted for a combined spinal epidural owing to our patient's difficult airway and past history of pulmonary tuberculosis. Different methods have evolved over the years to ease in identification of the intervertebral spaces like X ray of the spine. MRI and ultrasonography over the conventional landmark technique when encountered with a difficult spine. Each method has its limitations owing to non-feasibility in the operating room. Real time ultrasonography was found to be an easy, readily available technique in locating the most suitable intervertebral disc, the depth of insertion and the direction of Insertion of the Tuohy's and Quincke's needle [11,12]. Given the difficult anatomy in this patient we have justified the use of the ultrasound in identifying the appropriate space thereby alleviating agony to the patient in terms of multiple attempts and long durations of induction.

Conclusion

A case of successful spinal and epidural anesthesia in an

achondroplastic dwarf using ultrasonography to identify the space followed by loss of resistance technique to reach the epidural space for an orthopaedic procedure has been described. The use of real time ultrasonography in attempting central neuraxial block in cases wherein difficulty arises with the use of the traditional landmark technique is highly recommended.

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