Case Report

Achondroplasia - How do I manage this? A case report and review of the literature

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ABSTRACT

Achondroplasia, while being the most common form of disproportionate short stature, is a relatively rare case to encounter during anesthetic practice. With various problems, such as bony anomalies of the vertebral spine and difficult airway, planning anesthetic management of these patients requires special considerations. We report the case of a 54-year-old female with Achondroplasia, with fracture of the neck of the right femur treated by open reduction and internal fixation of the fracture. An appropriate sensory level could not be achieved on administration of spinal anesthesia despite an adequate dosage of Bupivacaine. Subsequently, conversion to general anesthesia was required. Surgery was uneventful, and the patient was discharged from the hospital after 48 h.

Key words: Achondroplasia, Difficult airway, Spinal anesthesia failure

Achondroplasia is the most common form of short-limbed dwarfism due to the FGFR3 mutation, with an incidence of 1 in 15000–30000 live births [1,2]. It is an autosomal dominant disorder that results in abnormal cartilage formation at epiphyseal growth plates. Due to various skeletal abnormalities, anesthetic management of these patients comes with its own set of challenges. There is limited literature regarding the failure of spinal anesthesia and subsequent conversion to general anesthesia in adults with achondroplasia, particularly concerning dosing considerations and intraoperative decision-making.

This case highlights the limitations of conventional height-based spinal anesthetic dosing and emphasizes the importance of individualized planning rather than relying solely on standard protocols.

CASE REPORT

A 54-year-old female presented with a history of a fall one day before admission to the hospital, with a fracture of the neck of the right femur.

Hemodynamically, the patient was stable. On examination, the pulse rate was 85/min, the respiratory rate was 18/min, and the blood pressure was 110/70 mmHg. There was no pallor, icterus, cyanosis, or generalized edema. On local examination, swelling and tenderness with restricted movement of the right hip joint were noted.

X-ray hip (Fig. 1) revealed a closed fracture of the neck of the right femur, with no distal neuromuscular deficit.

From an anesthetic perspective, metabolic equivalents were more than 4. The patient had short stature with a height of 128 cm, a weight of 30 kg, with a body mass index of 18.4 kg/m^2; upper segment: Lower segment ratio was 0.98, and arm span was 130 cm. Airway examination revealed adequate mouth opening with an inter-incisor distance of 4 cm and a Modified Mallampati grade of 2. However, the patient had a short neck with a sterno-mental distance of 10 cm and a thyromental distance of 4.5 cm. Dorso-lumbar kyphosis with narrowed intervertebral spaces was noted on spine examination and confirmed with spine X-ray (Fig. 2). No other abnormalities were noted on systemic examination. 2D Echocardiogram revealed mild mitral regurgitation, mild tricuspid regurgitation, with pulmonary artery systolic pressure of 18 mmHg and an ejection fraction of 60%.

Spinal anesthesia was attempted, but despite correct technique and appropriate administration of an adequate dose of Bupivacaine into the subarachnoid space, the sensory level could not be attained. Subsequently, general anesthesia was required to be administered. Propofol was used as an intravenous inducing agent. Airway was secured using No 3 Proseal due to the difficult airway, and sedation was maintained on oxygen, sevoflurane,

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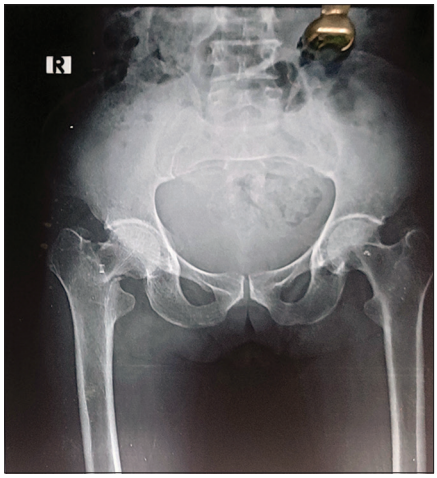

Figure 1: X-ray hip showing fracture of the neck of the right femur

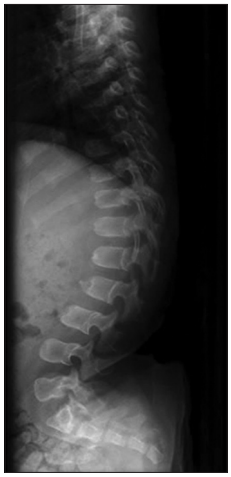

Figure 2: X-ray spine lateral view showing dorsolumbar kyphosis

and nitrous oxide. The duration of surgery was 2 h, after which the laryngeal mask airway was removed under deep plane of anesthesia, and the patient was monitored in the post-anesthetic care unit for 30 min. The patient was discharged from the hospital after 48 h of an uneventful stay.

DISCUSSION

Achondroplasia presents unique challenges to the anesthesiologist, particularly in the context of neuraxial and airway management. A fundamental dilemma in such patients is whether anesthetic planning should be guided by chronological age or by altered anthropometric parameters. While it is well-established that children are not "miniature adults," achondroplastic adults, conversely, exhibit anatomical characteristics, including shortened long bones, flat bone defects, large head size- frontal bossing, flattened nasal bridge, narrow foramen magnum, spinal lordosis, rhizomelic shortening, short metacarpals, brachydactyly, trident hand, varus leg deformity [3,4].

Certain factors make anesthetic management difficult, including a difficult airway, abnormal spine-scoliosis, kyphosis, and smaller vertebral spaces, which complicate the administration of neuraxial anesthesia, and unpredictable spread of anesthetic agent [4]. Other comorbidities, such as cardiovascular complications, obesity, add to the problems [5]. In addition, there is no data available on the appropriate dosing of anesthetic agents in these patients.

Neuraxial anesthesia in achondroplasia is inherently unpredictable due to several anatomical and physiological alterations [6]. These include spinal canal stenosis, reduced interpedicular distance, exaggerated lumbar lordosis, and possible epidural space narrowing [7,8]. Such variations can significantly affect the spread of intrathecal local anesthetic, often resulting in either inadequate block or unexpectedly high levels of anesthesia. Compounding this issue is the paucity of robust literature guiding optimal dosing of intrathecal agents in this population, with most evidence limited to case reports and small series [9].

In the present case, despite confirmation of correct needle placement by free flow of cerebrospinal fluid and administration of a dose calculated according to the patient's height, effective neuraxial blockade was not achieved. This failure may be attributed to erratic distribution of the anesthetic agent within a structurally abnormal subarachnoid space, leading to inadequate drug concentration at the intended neural targets [10,11]. It also raises the possibility that conventional height-based dosing paradigms may not be applicable in achondroplastic individuals, where body proportions are disproportionately altered.

The subsequent conversion to general anesthesia introduces an additional layer of complexity. Patients with achondroplasia are known to have difficult airway anatomy, including midface hypoplasia, macroglossia, limited neck extension, and potential atlantoaxial instability [12,13].

Furthermore, restrictive lung physiology and reduced functional residual capacity may predispose to rapid desaturation [14]. These factors necessitate meticulous pre-operative airway assessment and preparedness for difficult airway management.

This case underscores the limitations of relying on standard neuraxial techniques and dosing strategies in achondroplastic patients and highlights the importance of maintaining a low threshold for conversion to general anesthesia. A tailored, patient-specific approach, along with readiness to manage airway challenges, is essential to ensure perioperative safety in this unique population.

CONCLUSION

Achondroplastic patients present significant anesthetic challenges due to altered spinal anatomy and difficult airway considerations. This case highlights the unpredictability of neuraxial anesthesia and the limitations of conventional height-based dosing. Early recognition of block failure and timely conversion to general anesthesia are essential. Thorough pre-operative

assessment, airway preparedness, and a flexible and individualized anesthetic approach remain key to ensuring safe perioperative outcomes.

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