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ANAESTHETIC MANAGEMENT OF A 40-YEAR-OLD MALE WITH ACHONDROPLASIA UNDERGOING FEMUR ORIF: A CASE REPORT.

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Abstract

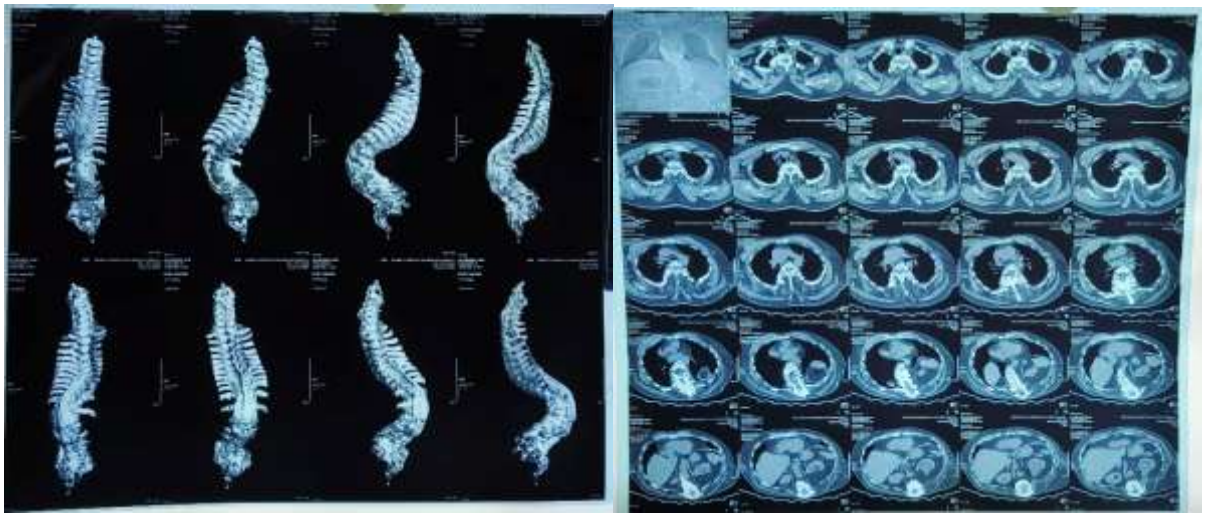
Achondroplasia is a genetic disorder characterized by skeletal dysplasia, airway abnormalities, and difficult neuraxial access, posing significant anesthetic challenges. Patients with achondroplasia commonly present with difficult intubation due to craniofacial abnormalities and may also have challenging neuraxial anesthesia due to spinal canal stenosis and abnormal vertebral anatomy. We present the case of a 40-year-old male with achondroplasia, scheduled for femur open reduction and internal fixation (ORIF). This report highlights the challenges encountered during airway management and the intraoperative approach to securing the airway under general anesthesia. A multidisciplinary strategy was crucial in ensuring safe perioperative management.

INTRODUCTION

Achondroplasia, the most common form of skeletal dysplasia, results from a mutation in the fibroblast growth factor receptor 3 (FGFR3) gene, leading to disproportionate short stature, macrocephaly, midface hypoplasia, and spinal abnormalities [1]. These anatomical variations present significant anesthetic concerns, including airway management difficulties, ventilation and potential perioperative respiratory complications [2]. Patients may exhibit cervicomedullary compression, foramen magnum stenosis, and thoracolumbar kyphosis, which complicate both regional and general anesthesia techniques [3].

Case Presentation

A 40-year-old male with achondroplasia was scheduled for open reduction and internal fixation (ORIF) of a femur fracture under general anesthesia. The patient had a history of short stature of Height – 125cms, macrocephaly, short neck with thyromental distance – 2.8cms , and limited neck extension, raising concerns about difficult airway management. Preoperative assessment revealed a Mallampati class IV airway, short thyromental distance, and restricted mouth opening, indicating a high likelihood of difficult intubation. The patient also had a kyphotic-scoliosis spine with suspected spinal stenosis, fused spinus process making neuraxial anesthesia technically challenging . Whole spine CT showed – fusion segmentation abnormality notes associated ankylosis of vertebral bodies and spinous process noted involving D12 dorsal and proximal visualised L1-3 lumbar vertebrae . Mild spinal cord narrowing noted in these levels . D10-L3 abnormal bone trabeculations noted . Dorso-lumbar kyphosis with scoliosis (C -shaped) spine



Inititally patient was planned for spinal anaesthesia and due to anatomical difficulties and unsuccessful attempts plan was made to go ahead with General anesthesia using Awake intubation . In the operating room , patient was prepared for Awake Fiberoptic Intubation by doing nasal packing using 10ml of lignocaine with adrenaline packing for 15 mins followed by topicalization of the airway with 4% nebulized lignocaine , 10% Lignocaine spray was sprayed on to the oropharynx , airway blocks (superior laryngeal nerve block using 2% lignocaine 10ml) were given and sedation with dexmedetomidine of 50mcg in 100 ml Normal Saline 20ml was given and initiated . Patient was initially preoxygenated and after adequate preoxygenation, awake fiberoptic intubation (AFOI) was performed following a 6.5 mm cuffed endotracheal tube was successfully placed with continuous visualization of the glottis using a fiberoptic Ambu scope . air entry was checked and EtCO2 was noted .

The patient was maintained on Pressure-controlled ventilation, with tidal volume delivering 375ml, Respiratory rate of 15breaths/min, PEEP of 4 , I:E 1:1.7 , Etco2 maintained at 30mmhg carefully adjusted to prevent barotrauma, given the restrictive pulmonary physiology associated with achondroplasia. Intraoperatively,

hemodynamic parameters remained stable, neuromuscular blockade was monitored using train-of-four (TOF) monitoring, and multimodal analgesia, including IV paracetamol and regional nerve blocks (femoral nerve block using 0.25% bupicaine 10 ml under USG guidance), was administered to minimize opioid use. The surgical procedure lasted 3 hours, with blood loss of 350ml (with in allowable blood loss limit) and controlled intraoperative fluid administration. At the end of surgery, the patient was transferred to the ICU for elective postoperative ventilation and monitoring, considering the risks of airway edema, residual neuromuscular blockade, and post-extubation airway obstruction. In the ICU, patient was on Fentanyl infusion of 4ml/hr for pain management and the patient was closely monitored for respiratory function, and after 6 hours, he demonstrated adequate spontaneous breathing, normal arterial blood gases, and was successfully extubated. Post-extubation, the patient remained hemodynamically stable, maintaining good oxygen saturation on supplemental oxygen, and was managed with multimodal analgesia and chest physiotherapy to optimize recovery.

DISCUSSION

Achondroplasia presents a variety of anesthetic challenges that require careful planning and execution to ensure patient safety. The most significant concerns in these patients are difficult airway management, difficulty in spinal anesthesia and perioperative respiratory complications. Given the skeletal dysplasia associated with achondroplasia, craniofacial abnormalities contribute to a high risk of difficult intubation, while spinal stenosis and vertebral malformations increase the likelihood of failed neuraxial anesthesia. This discussion elaborates on the various anesthetic considerations, particularly airway management, neuraxial anesthesia challenges, intraoperative ventilation strategies, and postoperative considerations, with a special focus on awake fiberoptic intubation and the Difficult Airway Society (DAS) guidelines for difficult airway management.

1. **Difficult Airway Management in Achondroplasia**

Patients with achondroplasia frequently present with macrocephaly, midface hypoplasia, a short thick neck, and restricted cervical spine mobility, which collectively contribute to a high risk of difficult mask ventilation and endotracheal intubation. The reduced mandibular size leads to a smaller oropharyngeal space, increasing the likelihood of airway obstruction under sedation or general anesthesia. Furthermore, atlanto-occipital instability and cervicomedullary compression may further limit head extension, increasing the complexity of intubation. The Mallampati score is often high (Class III-IV), thyromental distance is short, and the cervical spine is rigid, making direct laryngoscopy particularly challenging.

Given these structural abnormalities, awake fiberoptic intubation (AFOI) is considered the safest approach for securing the airway in patients with achondroplasia. AFOI offers several advantages, including maintaining spontaneous ventilation, avoiding excessive neck manipulation, and reducing the risk of airway collapse. In our case, fiberoptic intubation was successfully performed using a stepwise approach, beginning with adequate airway preparation using topical anesthesia (nebulized 4% lignocaine and 10% lignocaine spray), mild sedation with dexmedetomidine. The choice of dexmedetomidine over benzodiazepines or propofol was based on its ability to provide sedation without respiratory depression, allowing the patient to maintain a patent airway during intubation.

The Difficult Airway Society (DAS) guidelines emphasize that awake fiberoptic intubation should be the first-line approach in patients with anticipated Grade III or IV direct laryngoscopic views, severe craniofacial abnormalities, or limited cervical spine movement [1]. The DAS 2020 guidelines suggest a structured approach, including:

- Preoxygenation with 100% oxygen for 3-5 minutes to avoid hypoxia during intubation attempts.
- Optimal patient positioning, which, in achondroplasia, includes ramped positioning to align the oral, pharyngeal, and laryngeal axes.
- Adequate airway topicalization using nebulized or atomized lidocaine to prevent excessive coughing or airway reflexes.
- Stepwise advancement of the fiberoptic scope, ensuring a clear view of the glottis before tube advancement.

Following DAS recommendations, our approach included preoxygenation, sedation with dexmedetomidine, topical airway anesthesia, and careful fiberoptic scope advancement, leading to a successful intubation without complications.

2. **Challenges with Neuraxial Anesthesia in Achondroplasia**

While regional anesthesia is often preferred for lower limb surgeries, patients with achondroplasia frequently present with failed or difficult neuraxial anesthesia due to spinal canal stenosis, abnormal vertebral anatomy, and ossified ligamentous structures. In our case, attempts at spinal anesthesia were unsuccessful, suggesting spinal canal narrowing or altered epidural space anatomy.

Achondroplastic patients have a narrowed lumbar spinal canal, hypertrophic ligamentum flavum, and kyphoscoliosis, making both landmark-based and ultrasound-guided spinal anesthesia difficult. Studies have shown that in such patients, ultrasound guidance improves success rates by allowing for real-time identification of the spinal structures [2]. However, in cases of severe lumbar spinal stenosis, even with ultrasound guidance, the subarachnoid space may be inaccessible. Furthermore, epidural anesthesia is not always reliable, as epidural catheter placement may be challenging due to the presence of bony overgrowth and unpredictable spread of local anesthetic [3].

Given the high failure rate of spinal anesthesia in achondroplasia, general anesthesia is often required as an alternative. Preoperative discussions with the patient regarding the likelihood of failed regional anesthesia are essential in preparing for intraoperative anesthetic modifications. In our case, once spinal anesthesia was unsuccessful, the decision was made to proceed with general anesthesia with a secured airway.

3. **General Anesthesia Considerations in Achondroplasia**

Once the decision was made to proceed with general anesthesia, careful considerations were required to ensure safe intraoperative management. Pressure-controlled ventilation was preferred to avoid excessive airway pressures, particularly in patients with narrowed upper airways and restrictive pulmonary disease secondary to thoracic deformities. Ventilation strategies included tidal volumes of 6 mL/kg, a low respiratory rate, and PEEP pressure of 4mmhg to prevent air trapping , and barotrauma [4].

Another critical concern in achondroplastic patients is altered neuromuscular function, which can lead to unpredictable responses to neuromuscular blocking agents. Train-of-four (TOF) neuromuscular monitoring was used intraoperatively to ensure adequate muscle relaxation and to prevent overdosing of drug, reducing the risk of postoperative respiratory complications.

Fluid management was carefully titrated to prevent volume overload, as achondroplastic patients often have reduced vascular compliance, making them susceptible to fluid shifts and pulmonary edema. Crystalloids were administered at a controlled rate, and blood loss was minimal, decreasing the need for transfusion.

4**. Postoperative Considerations**

Patients with achondroplasia are at high risk for postoperative airway obstruction, especially after prolonged intubation and anesthesia. Factors contributing to this include macroglossia, pharyngeal narrowing, and potential airway edema following fiberoptic intubation. Therefore, the extubation plan must be carefully designed to prevent post-extubation respiratory distress.

In our case, extubation was performed in a semi-upright position to optimize upper airway patency. The patient was monitored closely in post-operative for signs of airway obstruction or hypoxia. Postoperative pain management included IV paracetamol 1gm TDS and regional nerve block (femoral nerve block using 0.25% bupicaine 10 ml under USG guidance) to reduce opioid use, minimizing the risk of respiratory depression [5].

CONCLUSION

The anesthetic management of patients with achondroplasia requires a multidisciplinary and highly individualized approach. Difficult airway management is the primary concern, necessitating careful preoperative planning, awake fiberoptic intubation, and adherence to DAS guidelines. Neuraxial anesthesia is often unsuccessful, requiring early discussion about alternative anesthetic plans. General anesthesia should be optimized with pressure controlled ventilation, neuromuscular monitoring, and judicious fluid administration. Postoperatively, patients must be monitored for airway obstruction, and a multimodal analgesia approach should be prioritized to reduce opioid-related complications. This case underscores the importance of anticipation, preparation, and a structured approach in managing patients with skeletal dysplasia undergoing orthopedic surgery.

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