Case study: Anaesthetic management of an achondroplastic dwarf undergoing radical nephrectomy

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Abstract
A three feet two inch (96 cm) tall achondroplastic patient with urothelial cell carcinoma involving renal pelvis was scheduled for a radical nephrectomy. Radial artery cannulation and central venous access were secured in the pre-induction period. After induction, the airway was secured using a flexible fibreoptic scope. General anaesthesia was maintained with oxygen-nitrous-oxide and continuous propofol infusion. The total duration of anaesthesia was three hours and 50 minutes. To the best of the authors' knowledge, this is the shortest adult achondroplastic patient ever reported to undergo such major abdominal surgery under general anaesthesia. The anaesthetic implications in patients with achondroplasia are reviewed in this case report.

Introduction
Dwarfism is defined as failure to achieve a height of 148 cm by adulthood.[1] More than a hundred different types of dwarfism have been described, but the commonest form of this relatively rare condition is achondroplastic dwarfism.[2] Anaesthetic management of achondroplastic patients undergoing varieties of surgical procedures – including limb lengthening surgeries,[3] Caesarean section,[4] bariatric surgery,[5] laparotomy for bilateral oophorectomy,[6] vesicovaginal fistula repair,[7] bilateral middle ear surgery and adenotonsillectomy[8] – have been reported. We describe the anaesthetic management of radical nephrectomy in a 96 cm tall achondroplastic dwarf with urothelial cell carcinoma involving the renal pelvis. This is possibly the smallest adult achondroplastic dwarf reported to undergo major upper abdominal surgery.

Case report
A sixty-five-year-old male achondroplastic dwarf with a right renal mass was scheduled for right radical nephrectomy. Initial pre-anaesthetic evaluation was done seven days prior to the surgery. He was a known hypertensive and had been on atenolol 25 mg for two months. Physical examination revealed a height of 3 feet 2 inches (96 cm) and weight of 54 kg. He had short limbs with a trunk of relatively normal length, protuberant head, saddle nose, large mandible and large tongue. Neck extension was normal with a Mallampati grade of I. The mentohyoid and mentothyroid distance were unremarkable. Cardiovascular examination was normal with blood pressure of 140/80 mmHg and pulse rate of 60/min. Respiratory examination was also not significant except for a depressed chest and protuberant abdomen with marked lumbar lordosis. Peripheral venous access was difficult because of stubby hands with coarse skin. Accurate non-invasive blood pressure measurement was also not possible due to short, stubby extremities. Breath holding time was 28 seconds and single breath count was greater than 30. Electrocardiogram showed incomplete right bundle branch block and left ventricular hypertrophy with q-waves in leads V1–V5. Transthoracic echocardiography showed concentric left ventricular hypertrophy, left ventricular diastolic dysfunction, ventricular ejection fraction of 60% and no regional wall motion abnormality. Chest x-ray showed more horizontally directed ribs with reduced intercostals distances. Pulmonary function tests could not be performed due to poor patient efforts. Contrast-enhancing computed tomography of the abdomen showed a large 10 x 10 x 8 cm well-circumscribed mass in the right kidney appearing to cross the adjacent right renal hilum and projecting to the aortic bifurcation and causing splaying of the renal pelvis around it with kinking of the inferior vena cava.

Incentive spirometry and deep breathing exercises were demonstrated and advised. The risk and benefits of epidural analgesia were explained in view of achondroplasia and major abdominal surgery. The patient, however, refused an epidural and opted for intravenous analgesia. The potential need for postoperative mechanical ventilation in a high dependency unit was also discussed with the patient. The patient was re-examined on the day prior to the surgery. Diazepam 5 mg and ranitidine 150 mg were prescribed on the night before and on the morning of the surgery. Antihypertensive therapy was continued on the day of the surgery.

After applying routine monitoring, venous access was secured using 22-gauge cannula in the right forearm after multiple failed attempts. Intravenous fentanyl 50 µg was administered for sedation and glycopyrrolate 0.2 mg for antisialagogue. Left radial artery cannulation was done under local anaesthesia after an initial failed attempt at right radial artery cannulation. Anatomical landmark-guided cannulation of the internal jugular vein (IJV) failed after repeated attempts because of poorly defined landmarks. Ultrasound was used to locate the IJV and cannulation was then done under local anaesthesia. A difficult airway cart including fibreoptic bronchoscope was kept ready. Anaesthesia was induced with intravenous propofol 100 mg and intravenous morphine 5 mg. Mask ventilation was possible and extreme care was taken to avoid hyperextension. Intravenous vecuronium 6 mg was administered for tracheal intubation. On laryngoscopy, a Cormack and Lehane grade III view was obtained and initial attempts at intubation resulted in oesophageal placement of the tracheal tube. Subsequently, the vocal cords were visualised using a fibreoptic bronchoscope and the trachea was intubated with a 7.5 mm cuffed PVC endotracheal tube. The endotracheal tube was fixed at 20 cm after checking the position with the fibreoptic scope. Anaesthesia was maintained with propofol with oxygen and nitrous oxide. Surgery was performed in the supine position using a transperitoneal approach. The intraoperative period was uneventful. Neuromuscular blockade was reversed with intravenous neostigmine 2.5 mg with glycopyrrolate 0.5 mg. Awake extubation was performed after adequacy of reversal was confirmed with a train-of-four ratio greater than 0.9, and the patient was making adequate ventilatory efforts. Postoperative pain was treated with tramadol. Incentive spirometry and deep breathing exercises were started in the early postoperative period.

Discussion

Achondroplasia is a rare (0.5–1.5/10 000 live births) genetic disorder characterised by abnormal endochondral bone formation. Spontaneous mutation accounts for 80% of cases, while the remaining 20% of cases are of autosomal dominant inheritance.[9] Point mutations in the human fibroblast growth factor receptor 3 (FGFR3) gene are well documented in inherited skeletal anomalies, such as achondroplasia and thanatophoric dysplasia, that are associated in most cases of dwarfism.[10] In addition, an oncogenic role has been proposed for mutant FGFR.[11] Recently, FGFR3 mutations were found in more than 40% of urothelial cell carcinoma patients.[12]

Anaesthetic management of an achondroplastic dwarf undergoing radical nephrectomy for urothelial cell carcinoma involving the renal pelvis has not been described so far. Both general and regional anaesthesia can pose problems in these patients. Difficulties associated with intubation in achondroplasia have been reported.[13–17] Short stature, enlarged head, saddle nose, maxillary hypoplasia, mandibular enlargement, megalocephaly with protuberant forehead and narrow nasal passages and nasopharynx are some features that contribute to airway difficulties in these patients.[14,15] Short neck leading to limited neck extension and marked cervical kyphosis with fusion of atlanto-occipital articulation may also be present. All these features may result in difficult mask holding/seal and direct laryngoscopy, with risk of forced extension leading to atlanto-axial dislocation.[13]

In our patient, difficult mask ventilation was not an issue but was considered due to the facial features described above. Muscle relaxant was administered only after ensuring adequate mask ventilation. No obvious airway difficulty was anticipated as our patient was Mallampati grade I with normal mentohyoid and mentothyroid distance, and normal neck extension. However, we encountered difficulty in visualisation of cords and a fibreoptic bronchoscope was used to secure the airway. As the glottic opening was small because of thickened cords, a smaller sized endotracheal tube was used. The length of insertion of tube was guided by fibreoptic scope to avoid any endobronchial intubation.

Short and stubby extremities make venous access difficult and the need for a central venous line should be anticipated in major surgeries. As difficult positioning and a short neck may have reduced the success rate, ultrasound-guided central venous cannulation was performed in our patient.

Cardio-respiratory functions may be impaired by several factors specific to achondroplasia. Rib hypoplasia with a flattened rib cage may result in reduced functional residual capacity and abnormal ventilatory mechanics.[13,14,15] Thoraco-lumbar kypho-scoliosis, rib deformities, upper respiratory tract obstructions and recurrent respiratory tract infections are common.[2] Obesity may further worsen the

pulmonary status of these patients.[18] Cor pulmonale is a relatively rare occurrence but has been reported as a consequence of these multiple respiratory complications.[2] Major upper abdominal surgeries in these patients may be associated with higher incidences of cardio-respiratory complications including pulmonary atelectasis, basilar consolidation, pleural effusion and V/Q mismatch, thereby resulting in prolonged mechanical ventilation. Postoperative pain, if present, may result in splinting and poor respiratory efforts. Bearing this in mind, consent for postoperative mechanical ventilation was obtained. Propofol was used for maintenance of anaesthesia to promote rapid awakening. If available, sevoflurane and desflurane could be equally beneficial. We succeeded in extubating the patient on the operating table after ensuring adequate muscle strength and ventilation along with good arousal.

Vertebral deformities, shortening of pedicles, decreased interpedicular distance and osteophyte formation make regional anaesthesia difficult in these cases.[19] Under-development of the vertebral arch leads to narrowing of the subarachnoid and epidural space.[16,17] A narrow epidural space makes dural puncture more likely, increases the difficulty in epidural catheter insertion and also limits the spread of local anaesthetics.[20] Further, there is increased risk of unidentified cerebrospinal fluid tap through epidural needle as the free flow of cerebrospinal fluid may be difficult to obtain. A high level of block is another complication reported with this technique.[3] Also, 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.[21] If necessary, small incremental doses of epidural local anaesthetics should be used to avoid complications. Due to these risk factors and the patient's refusal of epidural analgesia, tramadol was used judiciously for postoperative pain relief.

Conclusion

Anaesthesia poses a significant challenge to patients with achondroplasia. The need for meticulous assessment of anaesthetic risk in this group of patients must be recognised. Early communication with the anaesthetic team is important and a thorough preoperative assessment is mandatory. Facilities for managing the difficult airway and suitably experienced personnel should be available. Airway difficulty may be encountered even in the absence of known predictors. Fibreoptic intubation should be the technique of choice. Ultrasound-guided technique can be used to increase the success rate and decrease the complications associated with central venous cannulation. The risk of postoperative complications is high and facilities for postoperative mechanical ventilation should be arranged beforehand. The risk and benefits of epidural analgesia should be explained to the patient and alternative modes of pain control may need to be considered. Complete goal-directed workup and care of these patients will improve the outcome in this group of patients.

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