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Case Report

Anesthetic Management of Achondroplastic Dwarf with Difficult Airway for Cadaveric Renal Transplantation: A Case Report

Begüm Nemika Gökdemir*, Nedim Çekmen, Nazrin Zulfugarlı, Adnan Torgay

Department of Anesthesiology, Başkent University Ankara Hospital, Ankara, Turkey.

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ABSTRACT

Achondroplasia is a congenital autosomal dominant disease which has fibroblast growth factor receptor -3 mutation (FGFR-3). Anesthetic management of achondroplastic patients warrants special anatomical and physiological considerations because of important variations in the airway as well as the spine in regional techniques. In our case, 37-years-old male dwarf with achondroplasia has end stage renal disease (ESRD) and received cadaveric renal transplantation (RT). Preoperative assessment revealed the Mallampati score IV airway with a cervical instability, thoracolumbar kyphoscoliosis and spinal deformities. We aim to highlight anesthetic management of achondroplastic dwarf with difficult airway during operation.

Achondroplasia is a congenital disease which has FGFR-3 mutation and characterized with short limbs, midface hypoplasia, atlantoaxial instability. Achondroplasia is a genetic illness, and it has prevalence as 1:5000 to 1:40000 live births [1]. It is the most common form of dwarfism. There are many challenging factors such as accessing intravenous line, abnormality of skeletal, cardiopulmonary, central nervous system, craniofacial system, calculating drug doses and due to chondroplastic changes these patients have difficult airway and so its management is compelling for anesthesiologist [2-3]. Craniofacial deformities like large protruding forehead, big tongue, nasal, laryngeal hypoplasia and cervical abnormalities can cause difficult airway [3-4]. There are many medical complications associated with a diagnosis of achondroplasia. Increased vigilance is needed in both the evaluation and follow-up of this situation [2-3].

In this case, we discuss anesthetic management of an achondroplastic patient's difficult airway for cadaveric RT.

We obtained oral and written consent from the patient for the publication of this case report. This report adheres to the applicable guidelines of the Consensus-based Clinical Case Reporting Guideline Development in Enhancing the Quality and Transparency of Health Research.

Case Report

A 37-year-old male patient (31 kg, 123 cm) who has glomerulonephritis related ESRD, was admitted to our hospital for cadaveric RT. He had 3/7 days hemodialysis for 25 years and had fistula for this. At preoperative evaluation the patient was evaluated American Society of Anesthesiologists (ASA) III because of ESRD. He showed no signs of respiratory symptoms with exercise; however, he was not able to maintain a supine position for a long period because of shortness of breath and yet his pulmonary function test was normal. His preoperative laboratory results were between normal values except creatinine due to ESRD, his electrocardiogram and echocardiography were normal too. He also had achondroplasia with dwarfism, his neck was not flexible,

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*Corresponding author.
E-mail address: begokdemir@gmail.com

and he has cervical instability (Figure 1). His Mallampati score was IV (only hard palate was visionable). His teeth were protruded (Figure 2). Cervical thoracolumbar computer tomography (CT) and magnetic resonance imaging (MRI) showed that our patient has significant severe cervical instability, thoracolumbar kyphoscoliosis and spinal deformities. In Thorax CT, he has sclerosis appearances compatible with renal osteodystrophy at bone structures. There was a hypertrophic appearance in the thorax muscle structures and scattered focal emphysematous parenchyma areas in both lungs. We considered in preoperative evaluation room that he will be hard to intubate so we prepared difficult intubation cart which contains video laryngoscope, gum bougie, guide for endotracheal tube, different size laryngeal mask airways (LMA), endotracheal tubes, supraglottic airways and masks. And also, were placed in that cart and emergency cricothyrotomy set were ready in case intubation failed. The patient was admitted to the operation room where standard monitoring was instated including non-invasive blood pressure, electrocardiyogram, peripheral oxygen saturation, heart rate, end-tidal carbon dioxide, and body temperature. We inserted intravenous 20 Gauge cannula for vein access. Additional intravenous line was placed after multiple attempts. Anesthesia induction was maintained with propofol 2 mg/kg mg, lidocaine 0.5 mg/kg, fentanyl 2µcg/kg, rocuronium bromide 0.6 mg/kg. He used prednisolone 20 mg, so intraoperative we gave the patient 500 mg+500 mg pulse steroid. After 3 min preoxygenation, we intubated the patient with 7.0 endotracheal tube using videolaryngoscope on the third attempt (Figure 3).

After intubation, we initiated mechanical ventilation with 4-6 mL/kg tidal volume and maximum 4-6 cmH2O positive end expiratory pressure (PEEP) applied because he has emphysema. Anesthesia maintenance was provided by infusion of remifentanil 0.04-0.1 µcg/kg/min., rocuronium bromide 0.3-0.5 mg/kg/h and inhaler sevoflurane 2.2% in a 50% oxygen and air mixture. We performed serial arterial blood gas (ABG) monitoring in our patient and did not detect any acid-base disorder. The patient was given IL-2 antibodies and anti-edema treatment for RT. Fluid resuscitation was continued with Isolyte S due to normal potassium level. During the surgery, anastomoses were performed, and it was seen to be perfused by Doppler ultrasonography. No other complications developed. Since the incidence of pulmonary complications is high in achondroplastic patients, extubation was done with sugammadex 3 mg/kg and recruitment maneuver was performed in order to prevent postoperative atelectasis. Postoperatively, the patient was transferred to the intensive care unit. Immunosuppressive therapy was continued. He was taken to the ward after being in intensive care unit for 4 days. 8 days of staying in ward, he was discharged.

Figure 1- Posture of achondroplastic dwarf during preoxygenation

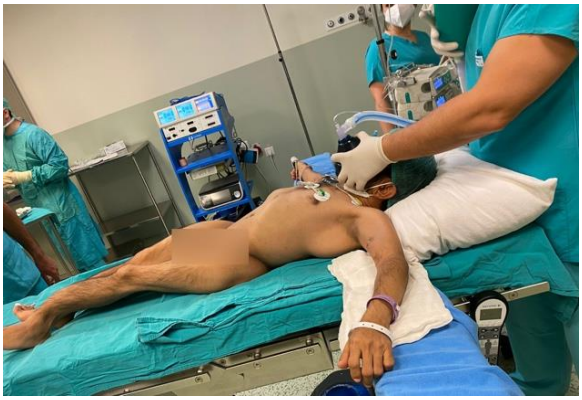

Figure 2- Protruded teeth, large mandible and large tongue

Figure 3- Intubation via video laryngoscope

Discussion

Achondroplasia is the most common type of dwarfism, and it contains multiple system abnormalities and comorbidities. Anesthetic management of achondroplastic patients warrants special anatomical and physiological considerations because of important variations in the airway as well as the spine in regional techniques. [2-3]

Those diagnosed with this disorder have increased mortality rates in childhood and adults.3 It can be hard for anesthesiologist due to difficult airway components. Preoperative evaluation should be multidisciplinary approach. In the light of literature, we prepared difficult intubation cart which contains video laryngoscope, gum bougie, guide for endotracheal tube, different size laryngeal mask airways (LMA), endotracheal tubes, supraglottic airways and masks. And also placed in the cart was an emergency cricothrotomy set in case intubation failed. In addition, calculating drug doses should be tailored ideal weight for height [5], so we can prevent overdose or underdose. The management of difficult airway should be planned for individual patients as a result of various deformities of craniofacial tract and multiple system abnormalities. To determine the risk of atlantoaxial subluxation secondary to odontoid hypoplasia and whether it is safe or not to perform traditional maneuvers for intubation, flexion-extension lateral cervical spine radiographs should be standard imaging [3-4].

If it provides inadequate imaging or there is a concern for cervical stenosis, CT or MRI scanning should be considered instead of radiography [6].

Cervical thoracolumbar CT and MRI showed that our patient had significant severe cervical instability, thoracolumbar kyphoscoliosis and spinal deformities. We were able to intubate the patient without complications, thanks to the availability of the difficult airway cart and preoperative preparation.

In this patient's had population, regional anesthesia can be applied as well as general anesthesia. However, since spinal deformities such as kyphoscoliosis and vertebral canal stenosis are common, the application of the technique becomes difficult [2-3-7].

We didn't prefer regional anesthesia because our patient has significant severe cervical instability, thoracolumbar kyphoscoliosis and spinal deformities. Despite of the anticipated difficulty in intubation and the fact that the patient came urgently for RT from a cadaver, it was decided to perform the operation under general anesthesia.

Another way of preserving the airway in such patients is awake intubation. The patient must be well informed for the procedure and preoperatively sedation is usually necessary. The cases performed by Nisa, et al. [7], and Kaushal, et al. [8] were performed as awake intubation because traditional airway maneuvers could have aggravated the risk of spinal injury. In our case, it was necessary to act quickly, since he came for emergency cadaveric RT. Spinal and thoracic cavity abnormalities can lead to restrictive lung disease, which is characterized with decreased functional residual capacity, atelectasis, increased closing volume and on long term obstructive sleep apnea, cor pulmonale and increase postoperative pneumonia. [2-9] Restrictive lung disease and pulmonary hypertension can prone to chronic hypoxemia or hypercarbia. Preoperative ABG may be warranted in these cases. [2-3-9]

We performed serial ABG monitoring in our patient and could not detect any acid-base disorder. To reduce pulmonary complications, we adjusted mechanic ventilation settings for individual patient and before extubation, recruitment maneuver was performed.

Conclusion

Achondroplastic patients with dwarfism are a challenging patient group for anesthesiologists in terms of existing systemic deformities as well as craniofacial and spinal deformities. We presented the case of cadaveric RT for achondroplastic patient with dwarfism and difficult airway. A multidisciplinary approach and comprehensive preoperative preparation is required in such cases.

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