CASE REPORT

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Anesthesia Management in an Acondroplasic Patient Developing Cardiac Tamponade Due to Myxedema

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ABSTRACT Skeletal dysplasia is a complex group of diseases that can occur with different clinical findings, and bone and cartilage tissue are affected. Various congenital malformations, spinal anomalies, limb shortenings and disproportionate length shortenings are seen in skeletal dysplasias, which can be divided into 40 main groups according to clinical, laboratory and radiological evaluations. In addition to bone anomalies, endocrinological problems such as hypoglycemia, hypothyroidism, hypocalcaemia, hypercalcemia, and adrenal insufficiency may occur. Achondroplasia is the most common type of skeletal dysplasia with autosomal dominant transition. There may be problems in anesthesia management due to limited neck extension, apnea and spinal anomalies. In this presentation, we aimed to present our general anesthesia experience in the female patient diagnosed with achondroplasia planned for pericardiocentesis due to cardiac tamponade in the light of the literature.

Keywords: Achondroplasia; general anesthesia; hypothyroidism; cardiac tamponade

Achondroplasia is the most common cause of dwarfism with short stature, short body and disproportionate development. This rare genetic disease, which is 0.5-1.5 in 10000 live births, is autosomal dominant inheritance and is common in women. Deficiency of endochondral bone formation and normal periostal bone formation disorder are characteristic; patients have bone deformities and systemic anomalies.[1] Atlantoaxial dislocation (AAD) may concurrently exist in achondroplastic patients either de novo, following surgery (foramen magnum decompression) or due to odontoid abnormalities (os odontoideum). Anesthetic management of achondroplastic patients with coexisting AAD offers a complex proposition for anesthesiologists in view of the anatomical and physiological constraints and the possible multisystem involvement.[2] In this case report, we aimed to present general anesthesia management in the context of the literature in an achondroplasic patient who was operated due to cardiac tamponade developed after pericardial effusion.

CASE REPORT

Pericardiocentesis was planned due to cardiac tamponade for a 24-year-old female patient, who is 92 cm tall and weighing 28 kg, diagnosed with achondroplasia in the ASA IIIE physiological class according to the American Society of Anesthesiologists (ASA). She was being treated at the endocrinology clinic due to myxedema and her T4: 0.9, TSH>100, with normal complete blood and renal functions. As she experienced respiratory distress, her endocrinologist demanded a cardiology consultation. Her echocardiography showed an ejection fraction of 60%, central arterial pressure 50 mmHg, tricuspid insufficiency, and a pericardial fluid surrounding the whole hearth. Her computed tomography revealed a malignant pericardial effusion entirely surrounding the heart, which reached 28-30 mm. She had distention due to abdominal ascites and her Glasgow Coma Score (GKS) was 12 (E:3M:5V:4). She had no limited neck extension, her mentohyoid and mentothy-

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roid distances were normal, and her Mallampati score was II. Auscultation revealed significantly reduced breath sounds bilaterally.

In the operating room, electrocardiography, peripheral oxygen saturation (SpO_2) and blood pressure (right radial artery) were performed, and her blood pressure before induction was 80/40 mmHg, cardiac apex beat 90/min, number of breaths 18/min and SpO_2 (on room air) 92%. Peripheral vascular access was provided with a 22-gauge venous catheter from the back of right hand after repeated unsuccessful attempts due to thickened skin tissue. No premedication was administered. In order to induce anesthesia, ketamin 2 mg/kg, fentanyl 2 mcg/kg and rocuronium 0.6 mg/kg were administered. Once adequate muscle relaxation was achieved, she was intubated using videolaryngoscope in a single attempt with number 5.5 cuffed endotracheal tube.

Ultrasound-guided right internal jugular venous cannulation was performed with a 4 Fr two-way central catheter. The maintenance of anesthesia was provided with 1% sevoflurane, 50-50% oxygen-air. In order to provide hemodynamic stability, 300 cc pericardial fluid was drained from the patient who needed noradrenaline infusion and no additional hemodynamic problems developed after drainage. During the 90-minute operation, the patient with a mean arterial pressure (MAP) in the range of 50-55 mmHg accompanied by a noradrenaline infusion was taken to intensive care in intensive care. In the postoperative period, analgesia requirement was achieved by ibuprofen medication (300 mg × 3 / day), in which sedation was provided with dexmetedomidine infusion (0.5 µg/kg/h) and noradrenaline infusion continued. The patient, whose cardiac and respiratory safety was provided, was extubated at the postoperative 65th hour. The patient, who did not experience any additional problems after extubation was transferred to the service at 72 hours. Informed consent was obtained from the patient's relatives that the clinical status of the patient in our case would be shared in a scientific journal.

DISCUSSION

Achondroplasia is a common non-fatal skeletal dysplasia. While autosomal dominant inheritance is observed in 20% of cases, spontaneous mutation is responsible in 80% of cases.[3] The point mutation in the Fibroblast Growth Factor Receptor 3 (FGFR 3) gene is responsible for development of hereditary skeletal anomalies such as achondroplasia and tanatrophic dysplasia, which are associated with most cases of dwarfism.[4]

The anesthesia management of patients with achondroplasia is complicated due to difficult airway. Short stature, large head and tongue, saddle nose, maxillary hypoplasia, mandibular enlargement, megalocephaly combined with protruding forehead and narrow nose are anatomical features that contribute to difficult airway.[5] There is limited neck extension due to short neck and atlantoccipital joint fusion combined with significant cervical kyphosis. All these features may be the cause of difficult airway and laryngoscopy associated with the risk of atlantoaxial dislocation, which may occur as a result of forced extension.[6] Smaller diameter endotracheal tubes should be preferred because of the thickened vocal cords.[7] In our patient, we did not have any difficulty in mask ventilation; although there was no evidence to suggest a difficult airway, we used videolaryngoscope for intubation for airway safety.

Short and stump extremities cause difficulty in providing peripheral vascular access and central venous access is recommended in major surgeries in these patients. Difficulty in positioning and short neck reduce the rate of successful cannulation and therefore ultrasound-guided central venous catheterization is recommended.[7]

Cardiorespiratory function is impaired due to many factors specific to achondroplasia. Thoracolumbar kyphoscoliosis, rib deformities, upper airway obstruction and related infections are common.[2] Short ribs and flattening of the rib cage cause functional residual capacity and abnormal respiratory mechanics.[5,6] Pulmonary hypertension and cor pulmonale are rarely seen in the presence of respiratory anomalies. Ventilatory insufficiency precipitation is possible due to anatomical and functional abnormalities of the rib cage. These patients are more prone to pulmonary complications such as atelectasis and pneumonia. They should be provided with ventilation

support until airway safety is ensured in the postoperative period.[2] Obesity exacerbates existing pulmonary problems in these patients.[8] Classification of the World Health Organization on obesity BMI 30.0-34.9 kg / m² (Class I); BMI is 35.0-39.9 kg / m² (Class II); BMI is 40 kg / m² and above (Class III or morbid obesity).[9] In line with this classification, our patient was evaluated as Class I obese. The intubation decision of the patient, who was intubated during the period to ensure stability in the cardiac and respiratory parameters in the intensive care unit, was made in accordance with the opinions of the surgeon and cardiologist who performed the operation. The intensive care follow-up period was also planned by the patient's surgeon and transplanted to the service after hemodynamic stabilization.

Whether general or regional anesthesia in achondroplasic patients is safe is a subject of discussion. Vertebral deformities, short pedicles, decreased interpedicular distance and osteophyte formation make regional anesthesia difficult. Inadequate development of the vertebral arch causes narrowing of the subarachnoid and epidural space. Narrowed epidural space is associated with difficulty in epidural catheter placement, limited local anesthetic distribution and increased risk of dural puncture.[7] High block level is among the reported complications.[10]

Childhood hypothyroidism causes delayed growth with delayed ossification and growth plate dysgenesis, while thyrotoxicosis accelerates ossification and growth. Thyroid hormone (T3) regulates chondrocyte proliferation and is required for hypertrophic differentiation. Fibroblast growth factors (FGFs) are also important regulators of chondrocyte proliferation and differentiation, and activating mutations of FGFR 3 cause achondroplasia.[11] A case report in which achondroplasia patient who developed myxedema was shared is available in the literature.[12] It has been confirmed by the cardiologist and endocrinologist that the pericardial and pleural effusion developed in our patient treated in the endocrinology clinic due to myxedema.

Dexmedetomidine is a lipophilic α2-metilol derivative with α2 adrenergic agonistic activity. It has sedative, analgesic and sympatholytic effects that suppress most of the cardiovascular responses seen in the perioperative period. Sedative and analgesic effects are mediated by α2 receptors in the brain (locus soreleus) and spinal cord. It reduces the need for intravenous and volatile anesthetics in intraoperative application, while reducing the need for combined analgesics and sedatives in the postoperative period.[13] In our patient, ibuprofen was adequate in analgesia management and we think that dexmetedomidine infusion contributed to this.

Anesthesia in achondroplasic patients is complicated due to differences in airway, cardiac, neurological and pulmonary functions. Care should be exercised during preoperative evaluation of these patients, while preparedness is required against any peroperative risks. Difficult intubation should always be kept in mind and safe conditions should be ensured. We believe that meticulous preoperative planning, intraoperative management and postoperative care support can help a surgical team achieve the desired results.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Sedef Gülçin Ural;* ***Design:*** *Sedef Gülçin Ural;****Control/Supervision:*** *Sedef Gülçin Ural;* ***Data Collection and/or Processing:*** *Sedef Gülçin Ural, Dilruba Güngör, Ömer Faruk Gülaştı;* ***Analysis and/or Interpretation:*** *Sedef Gülçin Ural, Dilruba Güngör;* ***Literature Review:*** *Sedef Gülçin Ural, Dilruba Güngör, Ömer Faruk Gülaştı;* ***Writing the Article:*** *Sedef Gülçin Ural;* ***Critical Review:*** *Sedef Gülçin Ural, Dilruba Güngör;* ***References and Fundings:*** *Dilruba Güngör, Ömer Faruk Gülaştı;* ***Materials:*** *Sedef Gülçin Ural, Dilruba Güngör, Ömer Faruk Gülaştı.*

REFERENCES

1. Ekwere IT, Edomwonyi NP, Imarengiaye CO. Anaesthetic challenges associated with achondroplasia: a case report. Afr J Reprod Health. 2010;14(2):149-55. [PubMed]
2. Kaushal A, Haldar R, Ambesh P. Anesthesia for an achondroplastic individual with coexisting atlantoaxial dislocation. Anesth Essays Res. 2015;9(3):443-6. [Crossref] [PubMed] [PMC]
3. Orioli IM, Castilla EE, Barbosa-Neto JG. The birth prevalence rates for the skeletal dysplasias. J Med Genet. 1986;23(4):328-32. [Crossref] [PubMed]
4. Vajo Z, Francomano CA, Wilkin DJ. The molecular and genetic basis of fibroblast growth factor receptor 3 disorders: the achondroplasia family of skeletal dysplasias, Muenke craniosynostosis, and Crouzon syndrome with acanthosis nigricans. Endocr Rev. 2000;21(1):23-39. [Crossref] [PubMed]
5. Morrow MJ, Black IH. Epidural anaesthesia for caesarean section in an achondroplastic dwarf. Br J Anaesth. 1998;81(4):619-21. [Crossref] [PubMed]
6. Mather JS. Impossible direct laryngoscopy in achondroplasia. A case report. Anaesthesia. 1966;21(2):244-8. [Crossref] [PubMed]
7. Jain A, Jain K, Makkar JK, Mangal K. Case study: anaesthetic management of an achondroplastic dwarf undergoing radical nephrectomy. S Afr J Anaesthesiol Analg. 2010;16(2):77-9. [Crossref]
8. Hecht JT, Hood OJ, Schwartz RJ, Hennessey JC, Bernhardt BA, Horton WA. Obesity in achondroplasia. Am J Med Genet. 1988;31(3):597-602. [Crossref] [PubMed]
9. Tam AA, Çakır B. Approach of obesity in primary health care. Ankara Medical Journal. 2012;12(1):37-41.
10. Brimacombe JR, Goddard JM. Leg lengthening in children-a retrospective review of anesthetic management in 61 children including 14 with achondroplasia. Paediatr Anesth. 2007;3(2):89-93. [Crossref]
11. Barnard JC, Williams AJ, Rabier B, Chassande O, Samarut J, Cheng SY, et al. Thyroid hormones regulate fibroblast growth factor receptor signaling during chondrogenesis. Endocrinology. 2005;146(12):5568-80. [Crossref] [PubMed]
12. Juarez M, Rohloff P. Myxoedema in a patient with achondroplasia in rural area of Guatemala. BMJ Case Rep. 2017;2017: bcr2016218506. [Crossref] [PubMed] [PMC]
13. Morgan GE, Mikhail MS, Murray MJ. Adrenergic agonists and antagonists. In: Butterworth J, Mackey D, Wasnick J, eds. Morgan and Mikhail's Clinical Anesthesiology. 5th ed. Lange medical book. New York: McGraw-Hill; 2013. p.239-53.