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Case Report

Anesthesia for tracheostomy in an infant with Apert syndrome*

Apert sendromlu bir bebekte trakeostomi için anestezi uygulaması

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Abstract
Apert syndrome is a rare, autosomal dominantly inherited disease characterized by irregular craniosynostosis and some malformations involving face, hands and feet. Respiratory functions are frequently deteriorated due to hypoplasia of oropharyngeal and nasopharyngeal cavities. Obstructive sleep apne syndrome, cor pulmonale and sudden death syndrome are among the complications of Apert syndrome. All of these anatomical and physiopathological disorders in the airways lead to a significant concern during anesthesia practice. Difficulty in mask ventilation, difficult intubation and postoperative airway obstruction may be seen in these patients. In this case report we present our anesthetic experience in an infant with Apert syndrome whom anesthesia was given for tracheostomy and difficult intubation was encountered.
Keywords: Apert syndrome; infant; general anesthesia

Özet
Apert sendromu, irregüler kraniosinostoz ile yüz, eller ve ayaklarda malformasyonlarla karakterize otozomal dominant geçişli nadir görülen bir hastalıktır. Orofarengeal ve nazofarengeal kavitelerin hipoplazisi nedeniyle, sıklıkla solunumsal fonksiyonlar bozulur. Obstrüktif uyku apne sendromu, kor pulmonale ve ani ölüm sendromu Apert sendromunun komplikasyonları arasındadır. Havayollarına ait tüm bu anatomik ve fizyopatolojik bozukluklar, anestezi uygulaması sırasında dikkatli olmayı gerektirir. Bu hastalarda maske ventilasyonunda zorluk, zor entübasyon ve postoperatif havayolu obstrüksiyonu görülebilir. Bu olgu sunumunda, trakeostomi nedeniyle anestezi uygulanan ve zor entübasyonla karşılaşılan Apert sendromlu bir bebekteki anestezi deneyimimizi sunuyoruz.
Anahtar kelimeler: Apert sendromu; bebek; genel anestezi

Introduction

Apert syndrome (acrocephalosyndactyly type 1) which is implicated among severe craniosynostosis syndromes was first described in 1906. It is characterized by some malformations involving skull, hands, feet and face. Craniosynostosis, midface hypoplasia and syndactyly of hands and feet are prominent features of this syndrome (1). Male and female sexes are equally affected with a prevelance of 12 per 1.000.000 live births (2). Although it is an inherited autosomal dominant disease, most cases are caused by new mutations (2). Musculoskeletal, cardiovascular, genitourinary and gastrointestinal abnormalities may also be associated with Apert syndrome (3).

Maintenance of airway is a major concern for anesthesiologists in the patients with Apert syndrome (1, 4). Ventilation by face mask may be difficult due to craniofacial malformations such as midface hypoplasia, choanal atresia and tracheal stenosis (5). Both facial abnormalities and limited neck movements may cause difficult intubation (5). Airway obstruction in the recovery period may be seen due to preexisting obstructive sleep apne syndrome (4). If airway obstruction was severe, invasive interventions such as tracheostomy may be required in these patients to maintain an open airway (6).

In this case report, we present a case of difficult intubation during anesthesia induction for tracheostomy in a patient with Apert syndrome.

Case

An infant who requires tracheostomy was referred to our hospital (Ankara Training and Research Hospital) from another hospital. She was born from a mother 34 years of age. She was the first live baby of her second pregnancy. Intrauterine growth retardation was diagnosed before birth whereas she was born with a birth weight of 2130 grams by caesarean section. The mother was receiving valproic acid for epilepsy and she could not get regular follow-ups during pregnancy. Any relation by lineal descent between parents as well as a history of similar malformed births in the family could not be detected. She was born premature with an Apgar score of 3 at 1 minute. Physical examination at birth has revealed intercostal retractions, atypical face appearance and respiration failure requiring ventilatory support. She was received continuous positive pressure (CPAP) therapy in intensive care unit. Two weeks later she underwent an operation due to diaphragma eventration. When she was 3 months-old, she was referred to

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otorhinolaryngology department in our hospital due to respiratory distress. The decision of otorhinolarygologists was to perform tracheostomy.

In the preoperative visit, the patient weighed 3040 grams and head circumference was 34 cm. She was suffering from a mild dyspnea while intercostal retractions were prominent. It was also noticed that she has triangular face appearance, broad fronthead, palpebral fissures, compressed nasal root, micrognathia and digital overlapping in feet. Laboratory tests including electrolytes, hepatic and renal function tests, electrocardiogram and pulmonary X-ray were normal.

On arrival at the operating room, monitoring including noninvasive arterial blood pressure, peripheral oxygen saturation, and electrocardiograms were applied along with preoxygenation. Various sizes of tracheal tubes, blades, laryngeal masks and stylets were prepared due to possibility of because of possible difficult intubation.

Anesthesia induction was provided by 8% sevoflurane in a mixture of 50% O_2/50% N_2O while ventilation by face mask was easy. After intravenous cannulation, 0.5 mg/kg rocuronium was administered to facilitate tracheal intubation. Oral intubation was attempted by 0 size Miller blade and size 3 uncuffed tracheal tube. Given the laryngoscopic view as Cormack-Lehane grade III, intubation could not be accomplished after three attempts. At the fourth attempt, intubation was accomplished by the aid of a stylet. After a failed insertion of a catheter via nasal route, gastric decompression was performed via oral route. Maintenance of anesthesia was provided by 2% sevofluran in the mixture of 50% O_2/50% N_2O. ETCO2 levels and anesthetic gas concentrations were also monitored throughout the operation. At the 60th minute, an incident of bradycardia (69 beats/min) was developed which was resolved by 0.02 mg/kg i.v. atropine. Duration of operation was 115 minutes and no other major complication was observed. SpO_2 levels were between 94% and 100% during surgery. At the end of the operation, the patient was transported to the intensive care unit while breathing spontaneously via tracheostomy tube.

Discussion

Apert syndrome is among the group of craniosynostosis syndromes. The most common reasons of operations of these patients are syndactyly and cranial malformations (5,7). Surgical interventions for respiratory disorders are also frequently performed, as in our patient. Because Apert syndrome patients may have anatomical airway malformations, anesthetists should be aware of respiratory complications such as difficult intubation and difficult ventilation (8, 9). Even if intubation was easy, intraoperative respiratory complications are not uncommon. Case studies has reported increase in secretion and wheezing during anesthesia in Apert syndrome (4,7-9). Increases in secretion may lead to desaturation and obstruction of tracheal tube (4,7). Studies has showed that one third of these patients experience intraoperative respiratory complications (4,8), whereas in 40% of them progressive airway obstruction has developed (6). The majority of intraoperative complications is related to supraglottic malformations and is encountered during induction of anesthesia (4). Fortunately, upper airway obstructions are easily resolved by simple airway maneuvers. Common causes of airway obstruction that are seen in Apert syndrome are midface hypoplasia, tonsillary hypertrophy, adenoid hypertrophy and choanal atresia (6).

It is also known that respiratory complications are common in patients with higher ASA (American Society of Anesthesiologists) scores and have an upper airway infection (8). Fusion of C5-6 cervical vertebrate which are seen in two thirds of Apert syndrome patients may yield to difficult intubations (5,10). So, laryngeal mask airway or face mask may be advantageous for short-duration minor operations (4,8). We did not experience any intraoperative respiratory complication. Although ventilation by face mask was easy, a difficult intubation was encountered.

In some cases, a nasal route could not be established due to midface hypoplasia in Apert syndrome (1). We could not insert catheter via nasal route, too.

Symmetric syndactyly is a common cause of surgery in Apert syndrome (5,7). Although syndactyly may also limit intravenous cannulation (5), we did not experience such a problem in our patient.

While musculoskeletal, and gastrointestinal abnormalities are frequently seen in Apert syndrome, cardiovascular and genitourinary abnormalities are not uncommon (3). The incidence of cardiovascular problems is 10% with its high mortality.

In conclusion, Apert syndrome which is characterized by severe airway and respiratory malformations needs great attention and careful preparation in order to decrease the risk of difficult intubation and cardiorespiratory complications.

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