Case Report / *Olgu Sunumu*

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Anesthetic Management of Patient for Case with Apert Syndrome

Apert Sendromlu Olguda Anestezi Yönetimi

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Abstract

Apert syndrome is an autosomal dominant inherited mandibulofacial dysostosis characterized by craniosynostosis, syndactyly, high forehead, broad nose, maxillary hypoplasia, synostosis of cervical vertebrae, organ malformations, and mental retardation. It is rarely encountered and as there is little knowledge of the anesthesia practice for this syndrome in the literature, we present our anesthesia experience of a case undergoing bilateral syndactyly surgery.

Keywords: Anesthesia, Apert syndrome, extremities-craniofacial abnormalities

Öz

Apert sendromu, kraniyosinostoz, sindaktili, yüksek alın, düz kemer burun, maksiller hipoplazi, servikal vertebralarda sinostoz, organ malformasyonları, konjenital kalp hastalıkları ve mental retardasyon ile karakterize ve sıklıkla otozomal dominant geçiş gösteren bir mandibulofasiyal dizostozdur. Nadir karşılaşılması ve literatürde bu sendroma ait anestezi pratiğinin az olması nedeniyle bilateral sindaktili ameliyatı yapılan olgudaki anestezi deneyimimiz sunulmuştur.

Anahtar Sözcükler: Anestezi, Apert sendromu, ekstremiteler-kafa yüz anormallikleri

Introduction

Apert syndrome (AS) is an autosomal dominant inherited mandibulofacial dysostosis characterized by craniosynostosis, syndactyly, high forehead, broad nose, maxillary hypoplasia, synostosis of cervical vertebrae, organ malformations, and mental retardation (1). The prevalence of AS has been reported to be 15/1.000.000 live births (2).

These patients have general anesthesia administered for a variety of operations and procedures in the pediatric period. We aimed to present our general anesthesia management for an AS case operated for bilateral syndactyly.

Case

Family consent was given for this case report. A 2-year-old, 10 kg female child operated for bilateral syndactyly was born by cesarean with 2020 g birth weight to a 31-year-old mother at the 34th week of the third pregnancy. It was learned that no one related to the mother and father or in the family had similar abnormalities. Intubated after birth, the patient underwent percutaneous endoscopic gastrostomy due to esophageal atresia at 10 days old, with surgery for bilateral choanal atresia at three months and isolated esophageal atresia at six months of age. After the operations, there was no need for intubation or mechanical ventilation and though admitted to hospital twice for pneumonia until two years of age, the patient had no problems with nutrition, snoring or sleep apnea.

Physical examination revealed reduced anterior posterior diameter and increased height of the head, flat and broad forehead, protruding temporal regions, flat occiput, proptosis, depressed nasal root, large ears, cleft palate, microphthalmia, enophthalmus, small mouth,

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micrognathia, and syndactyly of all extremities (Figure 1, 2). With preoperative consultations with ear nose and throat, neurosurgery and pediatrics requested, all other system examinations were normal. Mallampati score could not be assessed. Laboratory and echocardiographic investigations and abdominal ultrasonography identified no abnormalities.

Taken to the operating room without premedication, the patient had vascular access opened. The patient had standard (electrocardiography, peripheral oxygen saturation, non-invasive pressure measurements, and temperature) and bispectral index monitoring applied. Before general anesthesia, organization for possible difficult airway was completed. For induction intravenous (i.v.) 40 mg propofol, 10 mcg fentanyl and 10 mg lidocaine were administered. With no problems with mask ventilation, the patient was administered i.v. 5 mg rocuronium and was intubated with a number II Macintosh blade and 4.5 number cuffless tube on the first attempt. Anesthesia maintenance was ensured with 2.5-3% concentration of sevoflurane 50/50% oxygen/nitrogen protoxide and if necessary fentanyl and rocuronium. Intraoperative body temperature remained at normothermia. After the operation, i.v. 10 mg/kg paracetemol was administered for analgesia. During the 4.5 hour anesthesia, the patient did not develop any additional problems and hemodynamic parameters remained stable. With sufficient tidal volume reached, the patient was extubated when fully awake. After 45 minutes monitoring in the postoperative care unit, she was sent to the ward. The case developed no problem during follow-up and was discharged on the 8th day postoperative.

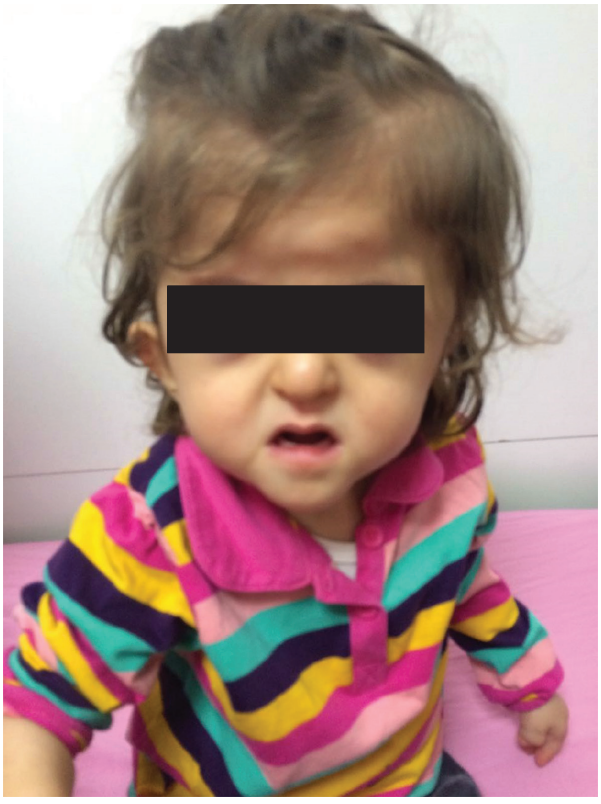

Figure 1. General body appearance with Apert syndrome

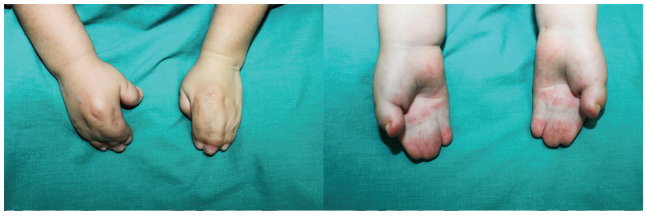

Figure 2. Bilateral syndactyly

Discussion

AS was first described by Eugene Apert(3) in 1906. The basic characteristic of the syndrome is premature fusion of the coronal and skull base sutures and agenesis of the sagittal suture leading to reduced nasopharyngeal diameter (4,5). The maxillary hypoplasia resulting from prevention of development of the maxilla in an anteroposterior direction causes stenosis of the nasopharyngeal airway and this deformity progresses with age (6). The leading airway abnormalities in AS cases are cervical vertebra fusion (71%), obstructive sleep apnea due to nasopharyngeal malformation and tracheal stenosis (bamboo trachea) linked to fusion of tracheal rings (7-9).

In these cases, preoperative problems, such as difficult airway, difficult venous access, airway hyperactivity, temperature irregularity, hemorrhage and venous air embolism risk with long surgery duration have been reported. The most significant concern has been stated to be difficulties ensuring reliable airway (10). The following may be listed among reasons causing difficulty ensuring reliable airway; angular deviation and strangulation of the trachea caused by disorders of cartilage tissue, tracheal ring fusion along with tracheal stenosis and shape disorders, limited neck movement caused by vertebral fusion of the cervical 5-6 vertebrae, structural disorders of teeth, cleft palate, and high palata (4,9).

Morris and Cooper (11) reported a 14-year-old patient with intubation difficulty who had undergone surgery under general anesthesia several times and had no intubation difficulty before. They reported that fibrosis and hardening of the temporal muscles developing in the maxillofacial region after surgical interventions may limit mouth opening with the most significant cause progressive traits of present structural disorders over time. As a result, for general anesthesia administration, they recommended preparations be made remembering that intubation difficulties may develop, even though their case had not

experienced difficulty securing the airway before. Due to abnormal facial characteristics of AS cases, appropriate choice of mask and intubation may be difficult. As a result, for our case, we prepared all necessary airway devices for difficult ventilation and intubation preoperatively. In spite of abnormal cranial and facial shape, we did not encounter any difficulty with ventilation and intubation.

Perioperative respiratory complications are commonly observed in AS cases. Complications that can be treated with bronchodilators or deepening anesthesia may occur, but may be severe enough to lead to cancellation in some cases. Development of respiratory complications is reported to be linked to accumulation of secretions caused by tracheal stenosis leading to a tendency for bronchospasm (5,9). We continuously monitored the airway pressure and had bronchodilators ready, but did not encounter any respiratory complications during or after the operation with our case.

With difficult venous access expected, we did not encounter this problem with our case. In our case, with cranial surface area increased due to craniosynostosis, warm fluids and heating blankets were used to preserve body temperature and prevent hypothermia. Additionally, as eyelids did not sufficiently close in this case, appropriate care was taken.

As a result, AS cases are candidates for frequent general anesthesia due to surgical interventions to correct present abnormalities. Due to abnormal airway, difficult ventilation, intubation and respiratory complications are expected during anesthesia. For reliable anesthesia management of these patients, we believe comprehensive evaluation and preparations for possible complications are required.

Author Contributions

Concept: G.K., B.A. Design: G.K., H.A., B.G.A. Data Collection or Processing: B.A. Analysis or Interpretation: H.A. Literature Search: G.K., B.A. Writing: G.K., H.A., B.GA. B.A.

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