Case Report / Olgu Sunusu

Artrogripozis Multipleks Konjenita ve Anestezi: Olgu Sunumu ve Literatürün Gözden Geçirilmesi
Arthrogrypozis Multiplex Congenita and Anesthesia: A Case Report and Review of the Literature

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Özet

Artrogripozis multipleks konjenita nadir görülen bir konjenital sendromu tanımlayan terimdir. Klinik tablo, tekrarlayan cerrahi girişimler gerektiren multipl eklem kontraktürleriyle karakterizedir. Çeşitli sistem anomalileri bu sendroma eşlik edebilir. Artrogripozis multipleks konjenitalı hastada anestezi yönetimi; zor havayolu, zor damaryolu ve malign hipertermiyi içeren birçok zorluğa yol açabilir. Biz bu olgu sunumunda, eklem kontraktürü için opere edilen 13 aylık erkek çocuğunun anestezi yönetimini tartışmayı amaçladık.

Anahtar kelimeler: Artrogripozis multipleks konjenita, anestezi, genel, hava yolu yönetimi

Abstract

Arthrogryposis Multiplex Congenita is a term that describes a rare congenital syndrome. Clinical features is characterized by multiple joint contractures that requires recurrent surgical interventions. Various system anomalies can be associated with this syndrome. The anesthesia management of a patient with Arthrogryposis Multiplex Congenita may lead to several challenges including difficult airway, difficult venous access and malign hyperthermia. In this case report, we aimed to present the anesthesia management of a 13 months-old case with arthrogryposis multiplex congenita who operated for joint contracture.

Keywords: Arthrogryposis multiplex congenita, anesthesia, general, airway management

Introduction

Arthrogryposis is a rare clinical condition characterized by non-progressive, congenital joint contractures in multiple areas of the body. This syndrome was described by Adolph Otto in 1841. Arthrogryposis term, literally meaning 'hook joint' in Greek, was used by Rosencranz. In 1897, Stern was defined the disorder as Arthrogrypozis Multiplex Congenita ([AMC], [1]).

The reported incidence of AMC in different studies varies from 1/12000 to 1/3000 live births. According to the animal studies, fetal akinesia leads to the congenital contractures, however the etiology has not been clearly known. Intrinsic and extrinsic factors such as olygohydramnios, hyperthermia, neurologic, muscular and connective tissue disorders, maternal illness and medications can cause to AMC (2).

Children with AMC often need several surgical procedures for treatment. In addition, difficult airway, difficulties with the insertion of a peripheral intravenous cannula, myopathy and risk of hyperthermia are the challenges for the anesthesiologist (3).

In this case report we aimed to present the anesthetic management of a patient with AMC.

Case

A 13-month-old boy of 6 kg weight with the diagnosis of AMC was scheduled for corrective orthopedic surgery. The patient had a medical history of recurrent pneumonia, two operations for congenital diaphragmatic hernia and cryptorchidism. In the first operation, the patient was successfully intubated in the third attempt by an experienced anesthesiologist, and a venous access was performed via internal jugular vein. In the third operation, laryngeal mask airway was used without any problem. Preoperative examination revealed multiple joint contractures involving upper and lower limbs, micrognathia, high-arched palate and short neck with an adequate neck extension and mouth opening. General anesthesia was planned, written informed consent was obtained for anesthesia.

After routine monitorisation, the induction of anesthesia was performed by using sevoflurane of 8%. The mask ventilation was easy. A 24 Gauge intravenous cannula was inserted to the dorsal surface of the left hand with difficulty. After a Cormack-Lehane grade II glottic view with direct laryngoscopy, anesthesia induction was completed with fentanyl 1 mcg/kg intravenous and rocuronium 0.6 mg/kg intravenous. An endotracheal tube of 4.0 mm sized was inserted easily at the first attempt of laryngoscopy. Anesthesia was maintained with sevoflurane, and mixture of 66% nitrous oxide and 33% oxygen. The temperature was monitored during the anesthesia and a convection warmer was used to avoid hypothermia. The surgery was performed in 60 minutes without any complication. The neuromuscular blockade was reversed with atropine 0,01 mg/kg intravenous and neostigmine 0,03 mg/kg intravenous. Morphine 0.05mg/kg intravenous was administered for postoperative analgesia. Thereafter, the patient was extubated, and taken to the recovery room. After an uneventful postoperative follow-up in the recovery room, the patient was admitted to the service.

Discussion

Arthrogrypozis Multiplex Congenita is a term consisting of several different clinical features. There is a large number of syndromes those can be associated with AMC. Respiratory problems, heart and kidney diseases can be developed due to lung hypoplasia, trachea-esophageal fistulas, congenital heart defects, and urogenital tract abnormalities (1,4). In preoperative period, examination of the organ systems is required for the quality and safety of anesthesia. In the present case, except history of recurrent pneumonia, all systems were normal. Thoracic radiography was performed for examining the respiratory system in preoperative period, and also pulseoxymeter

was used for monitorisation in the perioperative period

The etiology of AMC is still unclear. Various intrinsic and extrinsic factors, those lead to decreased fetal movement, impacts on muscle fibrosis, thickening and shortening of periarticular capsular tissue, and joint stiffness (1). In AMC patients, every joint is mostly involved. As a consequence of reduced oral aperture, high arched palate, mandibular hypoplasia, limited tongue, neck and temporomandibular mobility, cleft palate, short neck, torticollis and hemangiomas on the face, anesthesiologist have to be ready for difficult airway (4,5). The imaging of the cervical spine can be included in the preoperative assessment in case of limited neck mobility. Medical history can be beneficial for the possible risk of difficult intubation. In failed intubation, supraglottic airway devices, videolaryngoscopes, and airway exchange catheters can be the alternatives to the flexible fiberoptic bronchoscopes (6,7).

In the present case, also the patient's medical history revealed the successfully intubation, alternative devices for difficult airway management were kept ready. Tracheal intubation was performed on the first attempt uneventfully.

Furthermore, reduced subcutaneous tissue and muscle mass, contractures, osteoporosis due to lack of movement leads to difficulties in positioning and insertion of the intravenous cannula (1,8). Generally, AMC patient population is pediatric, and venous access can be unsuccessful. Therefore, inhalation agents are usually preferred for induction of anesthesia, which may lead to Malignant Hyperthermia (MH)—another challenge for the anesthesiologists. Relationship between MH and AMC is unproven. Several case reports showed that there was an increase in the body temperature and end-tidal CO_2 levels without any sign of cyanosis and myoglobinuria in the AMC patients during general anesthesia (4,9,10). Baines et al. (10) reported that no MH was detected in the 398 general anesthesia practice of 67 patients with the administration of halothane and succinylcholine. Otherwise, Baudendistel et al. (11) presented two cases of MH with AMC, in which the diagnosis was made with the muscle biopsy. Although most of the reported cases of MH are probably hypermetabolic reaction, it is recommended to avoid agents that can trigger MH. In addition, MH should be kept in mind in case of hyperthermia. Despite Chowdhuri et al. (9), who was described a case of hyperpyrexia in a six-day-old newborn during sevoflurane anesthesia, there has been numerous reports of uncomplicated anesthesia with sevoflurane (4). Due to anticipate difficult venous access we decided to inhalational anesthetic induction with sevoflurane. The patient's temperature was monitored carefully, all other resuscitation drugs and ice packs were kept ready. And also we avoided the succinylcholine because of the possibility of hyperkalemia and MH.

Spine deformity is seen in 10-30% of the patients with AMC (1). Despite of scoliosis and joint contractures those lead to difficulties in regional anesthesia, there has been few cases of AMC with successfully performed regional anesthesia (9). Regional anesthesia can be a good option in preventing difficult airway related complications, avoiding MH and relieving postoperative pain. Otherwise, opiates can be administered for postoperative pain therapy, however it is important to monitor the patient's respiratory functions postoperatively due to the possibility of respiratory depression (12). We prefer opioid for postoperative pain treatment and did not notice any adverse effects.

In conclusion, AMC lead to several anesthesia related concerns including malignant hyperthermia, difficulties in airway management, difficult intravenous access, and patient positioning. A careful preoperative assessment and appropriate perioperative monitoring are very important aspects of anesthesia in AMC to avoid complications.

Kaynaklar

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