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CLINICAL REPORT

Regional anesthesia with a single spinal anesthesia using hyperbaric bupivacaine in a child with arthroglyposis multiplex congenita

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Abstract Arthrogryposis multiplex congenita (AMC) consists of complex congenital anomalies characterized by multiple contractures. Anesthetic management of these patients requires special care: as this disease often progresses until dysfunction of multiple organ systems occur, it may have an impact on the anesthetic management. Here, we report a case of AMC undergoing urgent surgery for open tibia fracture who had difficult airway management because of limited mouth opening. The anesthetic management of this patient is represented in light of the literature.

Keywords Arthrogryposis multiplex congenita · Spinal anesthesia · Difficult airway management

Introduction

Arthrogryposis multiplex congenita (AMC) is an uncommon syndrome characterized by rigid joints and limb contractures that may be brought on by a variety of neurogenic, myogenic, skeletal, or environmental factors at birth [1]. Affected children often require anesthesia for surgical release of contractures and correction of associated defects. Anesthetic management of these patients requires special care [2]. Difficult tracheal intubation may be encountered because of limited neck extension, inadequate mouth opening, and short epiglottis [3]. Considering the difficulties related to the airway management, regional anesthesia may be an alternative, while it can create a challenge to the anesthesiologist.

In this case report, we describe the use of an anesthesia technique for urgent surgery for open tibia fracture in a child with AMC who has a history of difficult airway management. This procedure was performed under spinal anesthesia (SA) to avoid the need for tracheal intubation, which was predicted to be extremely difficult because of his temporomandibular joint (TMJ) contracture with limited mouth opening.

Case report

A 4-year-old boy weighing 15 kg with an open tibia fracture caused by trauma was admitted to our hospital. His physical examination revealed slight scoliosis, multiple joint contractures involving his limbs, and a TMJ contracture with markedly limited mouth opening (upper teeth to lower teeth, 1.3 cm) (Fig. 1). Respiratory and cardiovascular examinations were unremarkable. Laboratory testing included a normal serum creatine kinase level. A careful history revealed that the patient had a previous surgery for repair of club foot. During that operation, as intubation proved impossible despite several attempts by pediatric anesthesiologists experienced with both laryngoscopy and LMA, the case was ultimately managed with awake fiberoptic intubation at a pediatric hospital in another city. Because a fiberoptic bronchoscope suitable for children was not available in our clinic, we could not have used it for intubation during operation. After discussing possible options we planned to attempt a single-shot spinal anesthesia. We explained the planned anesthesia technique to his parents. After they understood and accepted this technique, a written informed consent was

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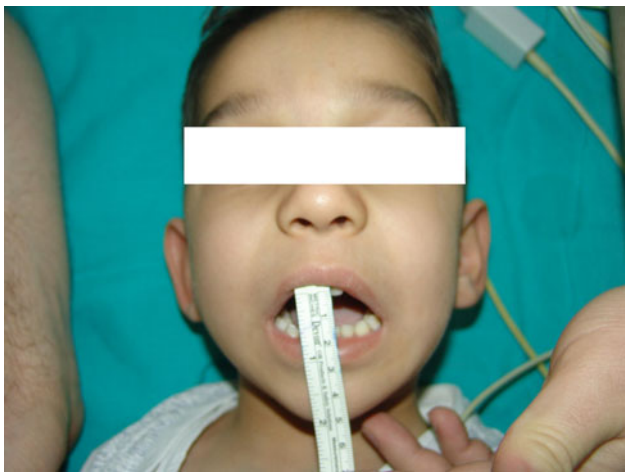

Fig. 1 A child diagnosed with arthrogryposis multiplex congenita presenting with markedly limited mouth opening

obtained. Because of difficult airway management, we called on a doctor from the Ear Nose Throat Department of our institution for urgent tracheostomy if needed. In the operating room, equipment for emergent tracheostomy were ready to use in case of a failed regional anesthesia or untoward local anesthetic reactions.

After placing routine monitors (ECG, NIBP, SpO_2), we administered oral ketamine 2 mg/kg with midazolam 0.2 mg/kg as premedication using a syringe. Then, a 25 gauge i.v. peripheral cannula was inserted after multiple attempts caused by the contractures. An infusion of Isolyte P (dextrose 5% with electrolytes: Na 25 mEq/l, Cl 22 mEq/l, K 20 mEq/l, Mg 3 mEq/l, P 3 mEq/l, lactate 23 mEq/l; Eczacıbası-Baxter, Istanbul, Turkey) was given at a rate of 5 ml/kg/h and continued throughout the operation. Approximately 5 min after sedation, the patient was placed in lateral decubitus position and alkalinized lidocaine was administered locally. After several attempts, the lumbar puncture was performed with a 90-mm 25 gauge Sprotte needle in the L4–L5 interspace. When the correct position of the needle was ensured [verified by free aspiration of cerebrospinal fluid (CSF)], 0.5% hyperbaric bupivacaine (Marcaine spinal heavy 0.5%; AstraZeneca, Wilmington, DE, USA) at a dose of 0.3 mg/kg with 1 μg/kg fentanyl was injected over nearly 10 s. During injection, the needle tip was positioned cephalad. He was then returned to the supine position. During surgery, heart rate was maintained at 85–100 beats/min and blood pressure at 60–90/35–55 mmHg. Oxygen 3 l/min was given via a pediatric nasal oxygen mask to maintain oxygen saturation at 96–100%. The dermatomal extension of the block and subsequent regression was determined by cutaneous finger pinching repeated every 5 min. Motor block was assessed with Bromage score. The highest level of sensory block was T5, and in the fifth minute Bromage score was 3. During the operation there was no need to give ephedrine and atropine to the patient. Nearly 70 min from the beginning of the surgery, the procedure was completed uneventfully. At the end of the surgery, 40 mg/kg rectal paracetamol was administered for postoperative pain. After the surgery, the patient was monitored in the PACU for at least 2 h, and when the motor block was completely resolved at 95 min, he was transferred to the surgical ward. His postoperative course was uncomplicated.

Discussion

Arthrogryposis multiplex congenita was first described by Otto in 1841 [4], and the term was created by Stern in 1923. The common primary disorder is deformed rigid joints [1]. Although the primary clinical manifestation is multiple joint contractures, there are often various associated anomalies involving the cardiovascular, nervous, genitourinary, and respiratory systems as well as airway abnormalities [1, 5]; the latter were seen in our patient. The most common maxillofacial findings reported include decreased mandibular opening, micrognathia, and high arched palate [6]. Of primary concern to the anesthesiologist is the potential for airway involvement, thereby making direct laryngoscopy and endotracheal intubation difficult [7]. Steinberg et al. [6] reviewed the findings in 23 patients with AMC and noted a 22% incidence of maxillofacial involvement with limited mouth opening in 3 of the 23 patients. The limited mouth opening in patients with AMC can be severe and may make direct laryngoscopy impossible, as in our patient [8]. In addition to these problems, these patients are prone to hypotension, respiratory depression, diminution of muscle tone, and prolonged action of various inhalational and intravenous anesthetic drugs [7]. There may be postoperative airway and respiratory issues with AMC. Scoliosis has been reported in up to 65% of patients with AMC [9, 10]. These problems also may result in alveolar hypoventilation, restrictive respiratory pattern, and postoperative atelectasis [7]. Patients with AMC may tend to show higher sensitivity to muscle relaxants [1]. Although increased risk for malignant hyperthermia has not been proven, several cases of hyperthermia have been reported in children with AMC [5]. Although regional anesthesia may have been difficult for these patients because of vertebral column deformities, in some cases spinal block was used for both caesarean section and ex utero intrapartum treatment in parturients with AMC [11, 12].

Our case was complicated by the fact that our patient had a known difficult airway during a previous operation because of AMC. He had a worse experience with the fiberoptic intubation; also, fiberoptic bronchoscope for children was not present in our clinic at this time. All these

considerations caused us to choose SA for this patient for urgent surgery.

In conclusion, we have presented our experience of SA in a child with AMC. Although this technique may be difficult because of vertebral abnormalities, it may be used as a reliable alternative to general anesthesia, especially at the facilities in which fiberoptic intubation is not available, as in our case. To our knowledge this is the first case in which SA has been used in a child with AMC. Considering the proposed urgent surgery and his known difficult airway management, we preferred SA, accompanied by preparations for emergency tracheostomy, if needed.

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