Kocaeli Med J 2018; 7; 2:23-25

Arnold Chiari Tip I Malformasyonlu Pediatrik Hastada Anestezi Deneyimimiz

Anesthesia Experience of a Pediatric Patient with Arnold Chiari Type I Malformation

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ÖZ

Arnold Chiari malformasyonu (ACM) konjenital olarak gelişen bir malformasyondur. Olguların bazılarına siringomiyeli de eşlik etmektedir. Dört tipi olup erişkinlerde sıklıkla tip I görülür. Tanı genellikle güçtür ve bundan dolayı gecikmiştir. Bu olgu sunumunda Siringomyelinin eşlik ettiği ACM Tip I nedeniyle opere edilen 4 yaşında bir hastada anestezi deneyimimiz sunulmuştur.

Anahtar Kelimeler: *Arnold Chiari Malformasyonu, Siringomiyeli, Genel Anestezi*

ABSTRACT

Arnold-Chiari malformation (ACM) is a congenital defect and some of the cases are accompanied by syringomyelia. There are four types of the malformation, type 1 being the most common in adults. Diagnosis of the malformation is usually difficult and therefore delayed. In this case report, we present our experience of anesthesia in a 4-year-old patient with ACM Type I with syringomyelia.

Keywords: *Arnold Chiari Malformation, Syringomyelia, General Anesthesia*

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Başvuru Tarihi: 26.01.2018
Kabul Tarihi: 13.05.2018

INTRODUCTION

Arnold Chiari Malformation (ACM) is a congenital anomaly defined as the displacement of the brain stem and cerebellum towards the cervical spinal canal. There are four types; Type I is the most common among adults (1). Syringomyelia was present in 30-70% of these cases (2). Airway management can be difficult for children with congenital malformation (3). This case report aims to present anesthesia management of a 4-year-old patient with Arnold-Chiari malformation type I with associated syringomyelia.

CASE REPORT

A 100 cm-long, four-year-old girl weighing 16 kg presented to the neurosurgery polyclinic with complaints of headache, gait disturbance, tip-toe and scissors gait and balance problems, pain in both arms, hypoesthesia below the knees in both legs and heat and cold sensitivity on hands and feet. Patient with ACM with cervicothoracic syringomyelia diagnosis was evaluated before surgery and was evaluated as (American Society of Anesthesiologists) ASA class II. Patient had no difficulty swallowing, physical examination revealed strength loss of 4/5 and she had no family history of ACM. Lhermite phenomenon (hyperextension of the body provoked by retroflexion of the head) was present.

In routine blood tests of the patient without cardiac pathology, Hb: 13.8, Hct: 41%, plt: 680,000, white blood cell count: 15.19, bleeding time 1.30 min, fasting blood glucose: 100, BUN: 15, creatinine: 0.5 mg / 139 mmol/l, K +: 4.8 mmol/l, INR: 0.92, aPTT: 29.7 and PT: 12.3 were measured.

The written informed consent was obtained after the parents were informed about the anesthesia and surgery. Patient was monitored with electrocardiography (ECG), non-invasive automated blood pressure (BP), pulse oxymetry (SpO2) and body temperature probe. BP levels at 101/53 mmHg, peak heart rate at 141 beats / min, and SpO2 at 99% were measured. Following a premedication of oral midazolam 2 mg, 30 minutes before patient was brought to the operating room, she was administered propofol 3.5 mg/kg, fentanyl 1.25 mcg/kg, rocuronium 0.6 mg/kg and 0.01 mg/kg atropine for anesthesia induction. She was intubated with a 2 mm spiral endotracheal tube on the second attempt. Hydration was achieved with 20 ml/h 1/3 isomix infusion through peripheral intravenous access with a 22G venous catheter on the dorsum of the right hand. Patient was given Concorde position and was supported with gel pads. Sevoflurane (2%) in oxygen/air (50-50%) mixture and 10 mcg fentanyl was used for anesthesia maintenance. Patient was hemodynamically stable during the 3-hour posterior fossa decompression with duraplasty and C1 posterior arch decompression operation. After extubation, patient was taken to the postoperative follow-up unit. Patient had blood pressure of 100/48 mmHg, heart rate of 128 beats/min and a saturation of SpO2: 98%. After a one-hour follow-up of stable hemodynamic status, the patient was transferred to the ward and after an uneventful postoperative recovery, she was discharged home 7 days after surgery.

DISCUSSION

ACM Type 1 includes a wide spectrum of symptoms and findings. 30% of all cases present no symptoms. Symptoms often are are silent and they vary a lot. Symptoms may include headache, paresthesia in all four extremities and cranial nerve dysfunction (4). Magnetic resonance imaging is the preferred imaging method to establish a definite diagnosis. ACM is accompanied by spinal and skull anomalies like basilar impression, atlanto occipital fusion, atlantoaxial assimilation, cervical spina bifida occulta, Klippel-Feil deformity, syringomyelia and scoliosis in %30-50 of the cases (1).

In a retrospective study of 31 pediatric patients under 6 years of age diagnosed with ACM Type I, the main complaints were impaired oropharyngeal function (35%), scoliosis (23%), headache or neck pain (23%), tactile sensitivity (6%) and strength loss (%3). Common physical findings were reported as abnormal tendon reflexes (68%), scoliosis (26%) and abnormal gag reflex (13%) (5). In addition, vocal cord dysfunction (26% in all patients under 3 years of age) and syringohydromyelia (52%) are also seen (5). In these patients kyphosis poses significant problems on airway safety. Both induction with inhalation agents leading to

hypercarbia and increased intracranial pressures; and the probability of respiratory depression and apnea caused by intravenous induction agents due to difficult airway, demands greater concern on anesthesia safety on patients with this diagnosis (6). A retrospective study of pregnant women with syringomyelia reports that general and neuraxial anesthesia can be successfully performed without major long-term complications (7). Unintentional dural puncture with epidural needle may lead to tentorial herniation, decreased perfusion pressure and herniation (8). The same complication may also occur with spinal anesthesia but the severity and frequency of dural puncture due to spinal needle size is less than epidural anesthesia. It has been emphasized that the choice of spinal and epidural needles in smaller sizes and avoidance of multiple interventions is important to improve anesthesia safety (8). Although there is no definite contraindication in the guidelines, general anesthesia is a good option because of increased intracranial pressure in ACM Type 1 patients (8). There is a report of two pediatric patients with no ACM type I diagnosis having unexplained sudden cardiopulmonary arrest resulting in death in the literature (9). There is also a case report in which ACM Type 1 and dolichoodontoid process are found to be responsible for this sudden and unexplained cardiopulmonary arrest (10). Risk of sudden death strengthens the need for ACM Type 1 to be evaluated in early surgical procedures.

The goal in anesthesia management of patients with ACM is to prevent the increase in craniospinal pressure and brain herniation. Extreme caution is necessary during anesthesia induction and extubation as coughing may cause an increase in intracranial pressure and consequently brain herniation. Tracheal intubation may be difficult or impossible to execute due to airway pathology, difficulty in positioning and limited neck movement. Since hypothermia may delay the return of postoperative spontaneous breathing, patients' body temperatures should be closely monitored during surgery.

REFERENCES

1. Rewari V, Singh S, Trikha A. Laparoscopic cholecystectomy in a morbidly obese patient with Chiari I malformation and craniocervical anomalies. J Clin Anesth 2008; 20:372-5.

2. Karavelioğlu E, Eser O, Boyacı MG, Çelik B. Arnold Chiari malformasyonu: Literatür eşliğinde vaka tartışması. Genel Tıp Derg. 2011; 21:29-34.

3. Isada T, Miwa T, Hiroki K, Fukuda S. The management of the difficult pediatric airway. Masui 2005; 54:490-5.

4. Chantigian RC, Koehn MA, Ramin KD, Warner MA. Chiari I malformation in parturients. J Clin Anesth 2002; 14:201-5.

5. Greenlee JD, Donovan KA, Hasan DM, Menezes AH. Chiari I malformation in the very young child: the spectrum of presentations and experience in 31 children under age 6 years. Pediatrics 2002; 110:1212-9.

6. Haldar R, Gyanesh P, Samanta S. Anesthesia for a patient of acromesomelic dysplasia with associated hydrocephalus, Arnold Chiari malformation and syringomyelia. J Anaesthesiol Clin Pharmacol 2013; 29: 555–7.

7. Garvey GP, Wasade VS, Murphy KE, Balki M. Anesthetic and Obstetric Management of Syringomyelia During Labor and Delivery: A Case Series and Systematic Review. Anesth Analg 2017; 125:913-24.

8. Choi CK, Tyagaraj K. Combined spinal-epidural analgesia for laboring parturient with Arnold-Chiari type I malformation: a case report and a review of the literature. Case Rep Anesthesiol 2013; 2013:512915.

9. Martinot A, Hue V, Leclerc F, Vallee L, Closset M, Pruvo JP. Sudden death revealing Chiari type 1 malformation in two children. Intensive Care Med 1993; 19:73-4.

10. Quebada PB, Duhaime AC. Chiari malformation Type I and a dolichoodontoid process responsible for sudden cardiorespiratory arrest. Case report. J Neurosurg 2005; 103:567-70.