Anesthetic management of a patient with Arnold–Chiari malformation type I with associated syringomyelia
—A case report—

Department of Anesthesiology and Pain Medicine, Wonkwang University Hospital, Iksan, Korea

Tai-Yo Kim, Cheol Lee, and Ji-Na Kim

Arnold-Chiari malformation type I (ACM I) is anatomically defined as the displacement of the cerebellar tonsils below the level of the foramen magnum. Syringomyelia is a condition in which a cavity called a syrinx develops in the spinal cord and is filled with cerebrospinal fluid. Here we report the anesthetic management of a case of ACM I with associated syringomyelia scheduled for suboccipital craniectomy, cervical laminectomy and duraplasty. (Anesth Pain Med 2012; 7: 166~169)

Key Words: Anesthetic management, Arnold-Chiari malformation type I, Syringomyelia.

Arnold-Chiari malformation type I (ACM I) is a disorder of uncertain origin that has been traditionally defined as tonsillar herniations of 3 to 5 mm or greater through the foramen magnum. The anomaly is a leading cause of syringomyelia and occurs in association with bony abnormalities at the cranio-vertebral junction [1-3]. Generally, ACM I and its associated disorders pose anesthetic risk; difficulty of airway management, autonomic dysfunction, avoidance of sudden increased intracranial pressure, and abnormal sensitivity to neuromuscular blocking agents. We present the anesthetic management of a case of ACM I and its associated syringomyelia scheduled for suboccipital craniectomy, cervical laminectomies and duraplasty.

CASE REPORT

A 34-year-old man patient weighing 68 kg was scheduled for suboccipital craniectomy, cervical laminectomies and duraplasty. He had been diagnosed with Arnold-Chiari malformation type I and its syringomyelia 10 years previously. Neurological symptoms including occipital headache, slurred speech, swallowing difficulty, weakness with atrophy in upper limbs, paresthesia of face, chest, and upper limbs, and loss of temperature and pain sensation had gradually progressed. Especially, occipital headache was aggravated by coughing, sneezing, or laughing. But balance and motor function were not affected. His cervical spine X-ray and computerized tomography (CT) (Fig. 1 and 2) showed bony fusion and deformity at the upper cervical spine and skull base. Magnetic resonance imaging (MRI) revealed downward herniation of the cerebellar tonsils below the foramen magnum, syringomyelia, fusion state of upper cervical spine and basilar invagination.

Fig. 1. Lateral cervical X-ray showed bony fusion and deformity at the upper cervical and skull base.
Fig. 2. Cervical sagittal (A) and coronal (L) CT with bone window settings showed severe deformity with basilar invagination and atlanto-axial assimilation.

Fig. 3. Sagittal T1-weighted MRI showed downward herniation of the cerebellar tonsils through the foramen magnum suggesting ACM I and syrinx within the cervical cord along the C2 and C3.

Anesthesiologists and neurosurgeons discussed the possibility to increase intracranial pressure with anesthetic and surgical procedures. We decided not to perform intracranial pressure monitoring in this patient because corrective surgery for this patient was supposed to decompress increased intracranial pressure accompanied by ACM.

Routine preoperative investigations were within normal limits. Airway examinations displayed an interincisor distance of 3 cm, Mallampati class III, thyromental distance of 6 cm, and severe limitation of cervical spine mobilization. Preoperative electrocardiography (ECG) showed sinus arrhythmia and chest X-ray was unremarkable.

In the operation room, the patient was monitored with pulse oximetry, invasive radial arterial and non-invasive automated blood pressure, capnography, bispectral index (BIS) and esophageal temperature probe. Muscle relaxation was monitored with peripheral nerve stimulator. There were anticipated difficulties with airway management in airway examinations. We therefore decided to perform nasal intubation under fiberoptic bronchoscopic control prior to surgery; this would be done with the patient awake. After intravenous midazolam 3 mg and a slow (60 seconds) bolus dose of remifentanil 0.5 μg/kg and followed by remifentanil infusion 0.15 μg/kg/min, the instrument was passed and lidocaine 10% sprayed under direct vision, via a small nylon catheter, onto and through the vocal cords. A well-greased and warmed 7.00 mm cuffed tracheal tube was then threaded over fiberoptic bronchoscopy which was introduced into the right nasal passage and both tube and scope were advanced to negotiate the corner into the pharynx. The scope was then maneuvered so that its tip passed through the vocal cords and the tube was passed into the trachea using the scope as a guide. The position of the tube was checked visually by reference to the carina, and by auscultation and confirmed by capnography.

Following tracheal intubation, anesthesia was induced with propofol 2 mg/kg and maintained with a mixture of oxygen/air (FiO₂ 0.5) and intravenous infusion of propofol and remifen-
tamil. Propofol infusion was titrated to maintain a BIS index of 40–60, a remifentanil infusion was titrated to maintain heart rate and arterial blood pressure within 20% compared with baseline values. Rocuronium 40 mg was given for proper muscle relaxation. Normocapnia was maintained throughout the procedure with a peak airway pressure 25 mmHg. The patient was covered by warming blanket to maintain normal body temperature during surgery. There were no significant changes in the hemodynamic status and body temperature of the patient until the end of surgery. Neuromuscular blockade was reversed with pyridostigmine 0.2 mg/kg, glycopyrrolate 0.008 mg/kg when the TOF ratio had returned to 25% at the end of surgery. When BIS values reached 80 and spontaneous breathing was achieved, extubation was performed. The patient was administered analgesics by patient-controlled analgesia pump containing morphine (60 mg), ketorolac (180 mg), and ramosetron (0.6 mg) in normal saline in a total volume of 100 ml.

The patient was transferred to the neurosurgical intensive care unit where he was under close observation for a day after surgery. He was transferred to the general ward after stabilization. The patient’s postoperative recovery was uneventful and discharged home 14 days after surgery.

DISCUSSION

The prevalence of ACM I defined as tonsilar herniations of 3 to 5 mm or greater is estimated to be in the range of one per 1,000 to one per 5,000 individuals [1]. The incidence of symptomatic ACM I is less but unknown. Other conditions sometimes associated with ACM I include hydrocephalus, syringomyelia, abnormal spinal curvature, tethered spinal cord syndrome, and connective tissue disorders such as Ehlers-Danlos syndrome and Marfan syndrome [2]. The ACM I is associated with the various skeletal and CNS abnormalities including basilar impression, atlanto-occipital fusion, atlantoaxial assimilation, scoliosis, or syringomyelia. The signs and symptoms of ACM I can be divided into those caused by the compression of dural or neural structure by the displaced cerebellar tonsils and those related to the progressive development of syringomyelia. The patients with ACM I usually become symptomatic in the late teens. However, some may first display symptoms at a more advanced age, even in the presence of the syringomyelia [4,5]. Diagnosis of ACM I is made through a combination of patient history, neurological examination, and MRI. MRI is the diagnostic test of choice for ACM I, since it easily shows the tonsilar herniation as well as syringomyelia, which occurs in 20 to 30 percent of cases [6].

The patients with ACM I and syringomyelia scheduled for cranietomy already present with some degree of neurologic involvement, which indicates significant compression of the neural elements in the cranio cervical junction. In patients with ACM I and syringomyelia, there is no evidence that any particular anesthetic agents are contraindicated and the proper anesthetic management has not yet been established. However, anesthetic management should begin with careful investigation of the medical history and a complete physical examination of the patient’s airway, respiratory, cardiovascular, and neurologic systems to exclude possible associated comorbidity. Patients with these conditions can pose challenges to the anesthesiologists during induction, maintenance or emergence of anesthesia and positioning; 1) difficulty of airway management, 2) autonomic dysfunction, 3) avoidance of sudden increased intracranial pressure, and 4) abnormal sensitivity to neuromuscular blocking agents.

First, ACM I is associated with bony abnormalities including cervical spine fusion at the cranio-vertebral junction, which lead to limited range of motion. Flexion-extension of the neck should be limited to prevent further compression of the neural structure. Airtraq™ laryngoscope or fiberoptic bronchoscopic intubation instead of standard laryngoscope is recommended in patients with skeletal cervical spine abnormalities and unstable cervical spine [2,7]. Careful planning for extubation, and possibly postoperative respiratory support with slow weaning, is indicated in patients with pronounced brainstem compression and cranial nerve involvement, owing to increased risk of postoperative ventilator failure [8] or compromised upper airway reflexes [9]. Our patient had severe limitation of cervical spine mobilization due to deformity and cervical spine fusion at the cranio-vertebral junction and therefore was intubated via flexible fiberoptic bronchoscopy from the start. After surgery, the patient was transferred to the neurosurgical intensive care unit where he was under close observation for a day.

Second, autonomic function should be evaluated in patients with significant brainstem involvement. Subclinical autonomic dysfunction, a well recognized condition in ACM I, can result in unstable hemodynamics, lack of compensatory response to hypothermia, hypotension, hypoxia, and hypocarbia intraoperatively. Invasive arterial blood pressure is monitored for measuring continuous blood pressure and blood gases analysis and body temperature should be closely monitored especially if autonomic dysfunction is suspected. The patient should be
closely monitored in the immediate 24 hours following surgery because of the risk of sudden apnea, cardiac arrest in syringomyelia associated with autonomic dysfunction [10]. Our patient was covered with warming blanket and monitored core temperature via esophageal temperature probe to maintain body temperature. A remifentanil infusion, and invasive radial arterial blood pressure monitoring enable close control of hemodynamic parameters and effectively attenuated cardiovascular response to intubation, a response which could potentially stimulate progression of a syrinx.

Third, sudden increase of intracranial pressure caused by induction, intubation, positioning, or extubation lead to spinal cord damage. The chosen anesthetic technique must firstly aim to avoid elevating intracranial pressure or changing cranio-spinal pressure gradients [11]. Normocapnia should be maintained and high airway pressures avoided to prevent any rise in intracranial pressure. Intravenous anesthetics except ketamine are more advantageous compared with volatile anesthetics for decreasing intracranial pressure. We used propofol as an anesthetics of induction and maintenance in our patient. Normocapnia via capnography and analysis of arterial blood gas was maintained throughout the procedure with a peak airway pressure 25 mmHg.

Fourth, patients with syringomyelia have an increased sensitivity to neuromuscular blocking agents [2,9]. The presence of syringomyelia is determined even in patients without a clinical picture of myelopathy. Succinylcholine should not be used to facilitate rapid intubation of the trachea. It is associated with hyperkalemia when given to patients with denervated muscle. The presence and location of motor deficit is noted to avoid overdosing non-depolarizing muscle relaxants by monitoring blockade on denervated muscles. It is therefore important to monitor the neuromuscular junction throughout the surgery. In our patient with weakness and atrophy of upper limb, rocuronium was used with for proper muscle relaxation accompanied by monitoring of neuromuscular function. He had normal return to full muscle function following a standard induction dose of rocuronium.

In conclusion, ACM I and its associated disorders pose anesthetic risks and the proper anesthetic management has not yet been established. A safe perioperative management of patients with ACM I can be achieved by careful attention to the derangements that occur with the disease. Optimal patient outcome will be improved with an interdisciplinary team management including anesthesiology, neurology and neurosurgery services.

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REFERENCES