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Case Report

Anesthetic management during Cesarean section in a woman with residual Arnold–Chiari malformation Type I, cervical kyphosis, and syringomyelia

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Abstract

Background: Type I Arnold–Chiari malformation (ACM) has an adult onset and consists of a downward displacement of the cerebellar tonsils and the medulla through the foramen magnum. There is paucity of literature on the anesthetic management during pregnancy of residual ACM Type I, with cervical xyphosis and persistent syringomyelia.

Case Description: A 34-year-old woman with surgically corrected ACM Type I presented for Cesarean delivery. A recent MRI demonstrated worsening of cervical xyphosis after several laminectomies and residual syringomyelia besides syringopleural shunt. Awake fiberoptic intubation was performed under generous topical anesthesia to minimize head and neck movement during endotracheal intubation. We used a multimodal general anesthesia without neuromuscular blockade. The neck was maintained in a neutral position. Following delivery, the patient completely recovered in post-anesthesia care unit (PACU), with no headache and no exacerbation or worsening of neurological function.

Conclusions: The present case demonstrates that patients with partially corrected ACM, syringomyelia, cervical kyphosis, and difficult intubation undergoing Cesarean delivery require an interdisciplinary team approach, diligent preparation, and skilled physicians.

Key Words: Anesthetic management, Arnold–Chiari malformation, cervical xyphosis, pregnancy and Cesarean section, syringomyelia

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INTRODUCTION

There are four types of Arnold–Chiari malformation (ACM; types I–IV).[27] Type I ACM has an adult onset and consists of a downward displacement of the cerebellar tonsils and the medulla through the foramen magnum. Type I ACM may manifest with headaches, neck pain, and mild coordination problems, but most often it is asymptomatic and discovered incidentally on brain or cervical spine MRI scans.[5,25,27] Advances in neuroimaging and more frequent use of MRI showed that the actual incidence of the malformation is much higher than initially thought (approximately 0.6–0.7%).[16]

The anesthetic concerns in women with Type I ACM are related to an increase in cerebrospinal fluid (CSF) pressure associated with pregnancy and the second stage of labor,[17] or a differential effect between cranial and spinal CSF pressure.

The purpose of surgical treatment is to provide wide decompression and is accomplished by removing the suboccipital bone and doing laminectomy of C1 and C2 followed by duraplasty.[24] Once the surgery is completed, the posterior fossa volume expands and the cerebellar tonsils ascend.[8,24] The underlying syringomyelia usually resolves over time. In some cases, the placement of syringopleural and syringoperitoneal shunts is necessary in order to improve patient symptoms such as headache, numbness, weakness, and coordination difficulties.[8,24]

It is commonly perceived that once surgical decompression is completed, there will be no further concerns regarding CSF pressure or the differential effect between cranial and spinal CSF pressures. In other words, labor and delivery, and the use of regional analgesia/anesthesia can be used with only the usual and customary precautions implied for any obstetrical patient.[14]

Not much is written about anesthetic management in parturients with Type I ACM who have undergone surgical decompression and who continue to suffer from neurological deficits.[1,19] We are presenting a woman in full-term pregnancy with a partially corrected Type I ACM, cervical xyphosis, and syringomyelia and an anticipated difficult airway management.

CASE REPORT

A 34-year-old, gravida 2 para 1 parturient was scheduled for Cesarean section at 38 weeks of gestational age. At age 19, she presented with paresthesias, headaches, neck pain, and poor balance, and Type I ACM with an extensive syringomyelia from the cervical to the lumbar area. Over the next several years, she had undergone surgical treatments, including suboccipital craniectomy, duraplasty with fascia lata graft, and decompressive C1–C6 laminectomies with placement of a syringopleural shunt. As a consequence of the laminectomies, the patient had residual cervical xyphosis with limited extension and continued to have symptoms related to Type I ACM, such as headaches, neck pain, stiffness, poor balance, and bilateral paresthesias of the upper and lower extremities (left side worse than right). Her symptoms were worsened by straining, coughing, and neck movements. Due to her residual Type I ACM, her neurosurgeon recommended avoiding normal vaginal delivery. The preoperative anesthetic evaluation showed a Mallampati grade III airway, limited mouth opening to 4 cm, and limited neck movement, with an anticipated difficult airway intubation. Recent MRI imaging of the cervical spine demonstrated worsening of the post-laminectomy cervical kyphosis, spinal cord atrophy, and a residual syrinx. The spinal cord was deviated posteriorly, the MRI of the thoracic spine demonstrated a syrinx in the entire thoracic cord, and the MRI of the brain showed structures of the hindbrain protruding through the foramen magnum [Figures 1–3].

The anesthesiology team, in conjunction with the obstetrical team, decided to perform awake fiberoptic intubation and utilize a multimodal general anesthesia. The patient and her husband were informed of an anticipated difficult orotracheal intubation. On the day of surgery, the patient was brought to the operating room with fetal heart rate monitoring in place. She received 0.2 mg IV glycopyrrolate, famotidine 20 mg IV, and 30 ml Shol's solution pre-operatively. Standard American Society of Anesthesiologists (ASA) monitors

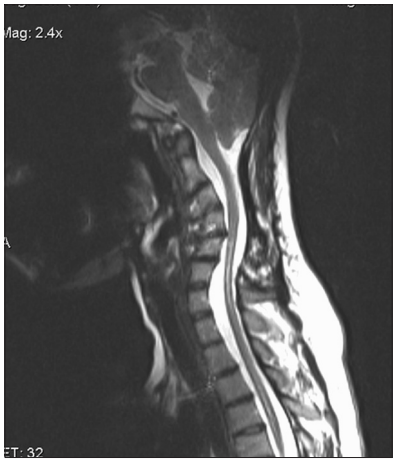

Figure 1: Sagittal T2 MRI image demonstrates severe post-laminectomy (C1–C6) cervical kyphosis (C2–C5), syringomyelia, and partially corrected Arnold–Chiari malformation Type I (tonsils below foramen magnum). Please notice crowding of the posterior fossa and anterior position of the larynx

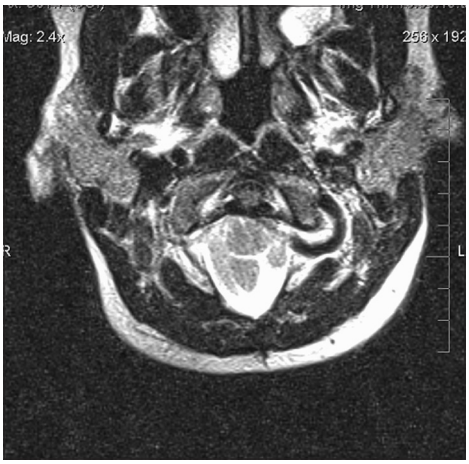

Figure 2: Axial T2 MRI image at craniocervical junction demonstrates cerebellar tonsils and syringomyelia

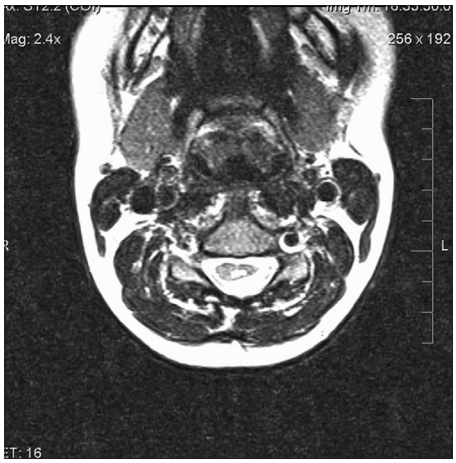

Figure 3: Sagittal T2 MRI image of thoracic spine demonstrates syringomyelia and atrophy of thoracic cord

and a Bispectral Index (BIS) monitor were applied. The neck was maintained in a neutral position. Adequate local anesthetic topicalization with Cetacaine (Cetylite Industries, Inc., Pennsauken, NJ, USA) topical oral spray (benzocaine 14%, butamben 2%, and tetracaine 2%) and 5 ml of lidocaine 2% jelly (Uro-Jet, International Medical Systems, Ltd., South El Monte, CA, USA) as a gargle solution were given in preparation of awake fiberoptic intubation. The patient received butorphanol 1.5 mg IV for sedation. Transtracheal 4% lidocaine 4 ml was administered. As the operative field was prepped and sterilely draped, the patient was properly prepared to undergo awake fiberoptic intubation with the head in the neutral position and with no neck movement. Utilizing an Ovassapian fiberoptic airway (Hudson RCI, Research Triangle Park, NC, USA), a pediatric bronchoscope (preloaded with a 6.5 endotracheal tube (ETT)) connected to an O_2 source was advanced into the trachea. It was noticed that the epiglottis was "floppy," angulated, the glottic opening kinked, and was cephalad and anterior due to the significant cervical kyphosis. No coughing or gagging was encountered, and the ETT was advanced to the supracarinal position and was secured.

General anesthesia with the neck in the neutral position was induced with propofol (2 mg/kg) IV as soon as proper ETT placement was confirmed by identifying breath sounds plus end-tidal CO_2 ($EtCO_2$) in addition to the fiberoptic visualization. Sevoflurane 2.0% end-tidal was initially used until the baby was delivered and then anesthesia was maintained with sevoflurane 1.0% end-tidal, plus an intravenous infusion of propofol (100 mcg/kg/min), midazolam 5 mg, and fentanyl (3 mcg/kg). A deep balanced anesthetic level was used without neuromuscular blockade. A healthy baby boy with Apgar scores at 1 and 5 min of 9/9 was delivered. The remainder of the case was without incident. The surgical team noted excellent uterine tone, and hemorrhage was less than average estimated blood loss ((EBL) < 700 ml). Post-intubation neurological assessment was performed and a prolonged in-house neurological observation was conducted. The patient had no complaints of headache or pain in the post-anesthesia care unit (PACU) or evidence of recall of the intraoperative events. She was discharged home after 4 days. Neurological exam showed no change in the patient's strength, sensation, or gross coordination. The patient remained stable at the outpatient clinic visit (2 weeks after Cesarean section), and she was advised to follow-up with her neurosurgeon with regards to progressive cervical kyphosis.

DISCUSSION

Anesthetic concerns in women with Type I ACM are related to an increased CSF pressure associated with pregnancy and labor, and also to the differential effect between cranial CSF pressure above the foramen magnum and the spinal CSF pressure below. This differential effect may worsen after lumbar tap or subarachnoid spinal blockade, which may lead to further descending of the tonsils with strangulation of the brainstem.[3,22,26] Syringomyelia associated with Type I ACM is a communicating type between the CSF and a syrinx and is thought to be related to compression of the subarachnoid space by the tonsils at the foramen magnum.[20] Anesthetic management for patients with syringomyelia is general anesthesia, avoiding CSF pressure fluctuation, and intracranial pressure (ICP) elevations without the use of spinal neuraxial anesthesia.[1,7,18] However, each anesthetic management plan has risks, and one "standard" plan is not appropriate for all patients.

The present case is a prime example of the individual anesthetic plan using a multidisciplinary approach. A full-term pregnant patient, with clinical residual Type I ACM, persistent cervical to lumbar syringomyelia, a syringopleural shunt, progressive cervical kyphosis, Mallampati III, and limited mouth opening with an interdental distance of 4 cm needed an anesthetic for operative delivery. The anesthetic plan in the present case was carefully formulated after all these factors were considered.

Since most anesthesiologists do not have the background or training to evaluate an MRI, it is important that they evaluate the MRI with a radiologist, neurologist, and/or neurosurgeon, even in patients with surgically treated ACM, with emphasis on the cerebellar tonsils and cervical cord prior to selecting the type of anesthesia.

It is commonly perceived that once surgical decompression is done, there will be no further concerns. Nonetheless, some patients have residual disease and the use of standard regional analgesic and anesthetic techniques in this situation, without the knowledge of

ongoing CSF pressure, may be harmful.[12,15] Incomplete decompression of Type I ACM may result after surgery with the development of new neurological conditions such as hydrocephalus, and progression of kyphosis after cervical laminectomies.[10,11] Post-laminectomy kyphosis is common in the cervical spine, especially when no augmented support, such as internal fixation, is used.[2] Progressive cervical kyphosis may lead to neck pain, gradual spinal cord compression, and myelopathy, which require further surgical decompressions.

General anesthesia without neuromuscular blockade and awake fiberoptic intubation were chosen in this patient to avoid CSF pressure fluctuation and ICP elevations. During the awake fiberoptic intubation, generous topical anesthesia was used to avoid coughing or bucking and changes in CSF pressures. It is also important to avoid neck movement and hyperextension during intubation in these patients, which may lead to further compression at the foramen magnum and cervical spinal cord, with traction on the lower cranial nerves and elevation of CSF pressure.

Patients with syringomyelia may be also sensitive to non-depolarizing neuromuscular blockade due to muscle wasting, atrophy, and hyperkalemic responses to succinylcholine administration.[9,28] However, proper muscle relaxation may prevent any movement and minimize the development of serious injury. Non-depolarizing muscle relaxants with judicious dosing and use of a neuromuscular twitch monitor as a guide may be an alternative in these patients. In addition, muscle relaxant reversal at the end of the case is recommended. It must be recognized that neostigmine could fail to reverse profound neuromuscular block in pregnant and post-partum patients.[6,23]

It is also important to prevent coughing and bucking during endotracheal extubation at the end of general anesthesia. Even in patients without hydrocephalus, increased ICP presents a risk for neurologic injury.[1] Extubation under deep anesthesia may decrease the risk of coughing, but it may increase the risk of gastric aspiration.

If neuraxial anesthesia is chosen for these patients, brain herniation may result. However, neuraxial anesthesia techniques are not contraindicated if a post-surgical MRI shows that the cerebellar tonsils ascend above the foramen magnum. Spinal neuraxial anesthesia performed in mothers who were found later to have uncorrected Type I ACM and syringomyelia was complicated by neurological worsening, persistent headaches,[13] nystagmus, and oscillopsia.[4] Signs and symptoms may develop up to 2 weeks after a dural puncture.[4,13] On the other hand, there are reports of successful neuraxial spinal anesthesia in women with surgically corrected Type I ACM.[14,17,21] There are case reports of epidural anesthesia done with injection of small local anesthetic bolus (3–4 ml) doses at 5-min intervals, with a total divided dose of 20 ml of 0.5% bupivacaine plus fentanyl 50 μg, with excellent results.[15,19] A spinal anesthetic using a 25-gauge needle and the injection of anesthetic into the spinal CSF (not the removal of CSF as done for diagnostic evaluation) may be another potential anesthetic plan for these patients.

We suggest that every patient having had ACM surgical correction should have at least a controlled MRI after surgery. If they do not, they need to have a consultation with a neurosurgeon, even if they are asymptomatic. Anesthesiologists thoughtfully consider whether using a spinal or epidural anesthesia in asymptomatic patients without consultation with neurology/neurosurgery is warranted. Sometimes, by asking targeted questions, physicians may discover symptoms of residual disease not disclosed by patients.

The present case shows that patients with partially corrected ACM, syringomyelia, cervical kyphosis, and difficult intubation undergoing Cesarean section require an interdisciplinary team approach, using diligent preparation and skilled physicians.

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