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Spinal neuraxial anaesthesia for caesarean section in a parturient with type I Arnold Chiari malformation and syringomyelia

Miqi Mavis Teo

Abstract

Introduction: Type I Arnold Chiari malformation is associated with prolapse of the cerebellar tonsils into or below the level of the foramen magnum and is usually diagnosed in adults. There are no current guidelines for the management of patients with a residual type I Arnold Chiari malformation, planned for a caesarean section under spinal neuraxial anaesthesia. The paucity in the literature on this topic presents as a management dilemma.

Case report: We report a case of a term parturient with type I Arnold Chiari malformation, following surgical decompression 4 years earlier, with a residual syringomyelia that underwent an elective caesarean section under spinal neuraxial anaesthesia.

Conclusion: This case highlights that multidisciplinary management and early anaesthetic consult are of paramount importance in the outcome of the patient, and that spinal neuraxial anaesthesia can be considered as a safe anaesthetic option.

Keywords

Arnold Chiari malformation, pregnancy and caesarean section, spinal neuraxial, anaesthetic management

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Introduction

Arnold Chiari malformation (ACM) can be divided into four types (types I–IV). Type I is a congenital neurological anomaly associated with prolapse of the cerebellar tonsils into or below the level of the foramen magnum and is usually diagnosed in adults and approximately 30%–50% of patients with type I ACM have an associated syringomyelia. They typically present with headaches, neck and shoulder pain, paraesthesia and mild incoordination, though most of them are asymptomatic and discovered incidentally on brain or cervical magnetic resonance imaging (MRI) scans.[1–3]

Suboccipital craniectomy and decompression has been of modest benefit in improving the symptoms caused by craniospinal pressure dissociation associated with the disordered flow of cerebrospinal fluid (CSF). The best results are obtained when surgery is performed on symptomatic patients or those with severe findings.

There have been case reports of the use of spinal neuraxial anaesthesia (NA) for caesarean sections in such patients, though the information is limited with no consensus on the topic thus far. We are presenting a parturient with type I ACM and syringomyelia planned for a caesarean section.

Case report

A 40-year-old female, height 145 cm, weight 45 kg, gravida 1 para 0, was planned for an elective caesarean section at 38 weeks of gestational age. She has a significant past medical history of thalassaemia minor, migraine and a type I ACM. She presented 5 years ago with persistent left-sided headaches, numbness of the left side of the face, upper limb and neck. The initial MRI had depicted a large syringohydromyelia seen in the entire spinal cord extending from C1 to T12 level with only a small syrinx in the conus medullaris. There was herniation of the cerebellar tonsils below the level of the foramen magnum by about 11 mm. She underwent a foramen magnum decompression. The intraoperative findings were that of a thickened dural band at the foramen magnum, with elongation of tonsils down to the upper border of C1. The follow-up MRI done the following year showed that

Division of Anaesthesiology and Perioperative Medicine, Singapore General Hospital, Singapore

Corresponding Author:
Miqi Mavis Teo, Division of Anaesthesiology and Perioperative Medicine, Singapore General Hospital, Outram Road, 169608 Singapore.
Email: mavis.teo.m.q@singhealth.com.sg

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the CSF space around the craniocervical junction had improved, though the large syrinx still persisted and extended from C1 to beyond T5.

Neurosurgical consult was made early in pregnancy by the obstetrician and she was deemed suitable to proceed with either a normal vaginal delivery or a caesarean section.

Pre-operative assessment was conducted 1 day prior to the surgery. There was residual left-sided numbness over her face and left upper limb and the headaches were still present but not worsening. There were no other signs or symptoms of raised intracranial pressure (ICP). These symptoms were not aggravated with pregnancy, coughing, or neck movements.

A physical examination was done, specifically, there was decreased sensation over the left face and left upper limb. Airway examination was unremarkable. There was a slight thoracic scoliosis over her back but the spinous processes were well felt in the lumbar region.

She was counselled regarding the risks and benefits of spinal NA versus general anaesthesia (GA) together with the obstetrician, with consideration of the input from the neurosurgeon. A normal vaginal delivery was discussed again as it was not a contraindication. However, she had concerns with lying supine for a prolonged period due to her thoracic scoliosis.

She had initially decided to proceed with the caesarean section under a GA. However, after thorough discussion of the risks and benefits of a GA versus spinal NA, the decision was made to proceed with the surgery under spinal NA.

The patient was positioned in the sitting position, and a single shot spinal anaesthetic was given at the level of L3/4 interspace. This was a single attempt using a 27-gauge Whitacre spinal needle, with a spinal introducer. Given her relatively short stature, intra-thecal hyperbaric bupivacaine 0.5% 1.6 mL with fentanyl 15 µg and morphine 100 µg was administered. Assessment of block height to cold sensation was performed, and a level corresponding to that of T4 was achieved prior to surgical incision.

Intra-operatively, the patient had mild discomfort from intraoperative surgical stretching of the abdomen, which was transient in nature. She was given intravenous (IV) fentanyl 50 µg in titrated boluses, together with Entonox, just prior to the delivery of the baby. After the delivery of the baby, she was given IV 1 mg of midazolam. The surgery was completed uneventfully thereafter without further complaints. Post-operatively, she was monitored in the high dependency unit overnight. Follow-up on the patient on the first day following surgery revealed no worsening of her pre-existing neurological symptoms, which was that of a left-sided numbness over her face and left upper limb. There was also no interval development of new neurological symptoms. She was discharged on post-operative day 3 and remained well on her follow-up visit with the obstetrician 2 weeks later.

Discussion

There is a lack of literature on the management of a pregnancy in a woman affected by ACM. There is little information regarding the safety of allowing labour or the use of NA in women with corrected ACM, and these anaesthetic practices have been restricted to case reports and one small retrospective case series.[4] An ACM with a concomitant syringomyelia implies that there is an initial or persisting continuity between the syrinx and CSF in the central canal of the cord. The foramen magnum abnormalities cause intermittent obstruction to CSF outflow from the fourth ventricle, with development of craniospinal pressure dissociation with a relatively higher CSF pressure in the head and a lower pressure in the spine.

Anaesthetic concerns in women with type I ACM are related to an increased CSF pressure associated with pregnancy and labour 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.[5–7] Traditionally, anaesthetic management of these patients would be a GA, avoidance of CSF pressure fluctuation and ICP elevations. Although there is a theoretical risk of herniation with sudden decompression of spinal column by NA, there is some evidence that active neurological disease is no longer considered an absolute contraindication to NA.[8]

This case highlights the importance of early multidisciplinary approach to a complex issue with an individualized plan. It is commonly perceived that once surgical decompression is carried out, there will be no further concerns. However, some patients may have residual disease and the use of standard regional analgesic and anaesthetic techniques in this situation, without any knowledge of the ongoing CSF pressure, may in fact be harmful. As most anaesthesiologists do not have the background or training to evaluate an MRI, it is important to evaluate the scans with a neurosurgeon/radiologist, with emphasis on the cerebellar tonsils and cervical cord, prior to anaesthetic selection.

In this case, the patient had opted for an elective caesarean section. She was offered a spinal NA. There are theoretical concerns with spinal anaesthesia in uncorrected or symptomatic patients with type I ACM because of the potential for compression of structures at the level of foramen magnum, increased ICP or obstructive hydrocephalus, and intramedullary cervical cord syndrome. There have been reports of worsening of symptoms up to 2 weeks after a spinal anaesthetic was administered. In two case reports, the ACM was unknown at the time of delivery and only diagnosed subsequently because of adverse neurological outcomes.[9,10] There have also been reports of successful NA in women with surgically corrected type I ACM. In these cases, there was active involvement of the neurosurgical team, and it was determined that a NA was not contraindicated because a dural puncture should neither impair CSF flux nor precipitate bulbar compression.[11,12]

We opted for a single shot spinal anaesthetic with a 27G Whitacre needle as our anaesthetic of choice. This would reduce the risk of local anaesthetic (LA) toxicity and avoid

the risk of an inadvertent dural puncture with an 18G Tuohy needle when attempting an epidural anaesthetic. A single shot spinal anaesthetic with hyperbaric bupivacaine and only fentanyl may be more beneficial in the detection of neurologic disturbances when compared with an epidural which requires repeated dosing. Furthermore, epidural anaesthesia might raise the ICP when a bolus of LA is given due to dural compression in the epidural space with a shift of CSF into the cranium.[13] However, slow and small increments of LA in the epidural space may circumvent this issue. A neuraxial technique may offer several advantages in that it avoids the potential hazards of securing the airway, respiratory function is less compromised and the existing craniospinal CSF pressure relationship is better preserved.

When considering a neuraxial technique, it is important to look for associated abnormalities such as kyphoscoliosis and spina bifida. Neurological assessment should include a check for evidence of autonomic neuropathy. Identification of autonomic neuropathy is paramount as these patients commonly present with tachyarrhythmias and widely fluctuating arterial pressures in response to stress and anaesthesia.[14] Sudden cardiac and respiratory arrests have also been attributed to autonomic neuropathy in patients with syringomyelia after posterior fossa surgery.

With signs of increased ICP, GA would be preferable over any type of NA. However, with the need to perform rapid-sequence induction, provide hyperventilation, avoid neck hyperextension that might further compress the brainstem, and because of the risk of possible vocal cord paralysis and stridor caused by traction on cranial nerves, GA for type I ACM with increased ICP can be particularly challenging.[15]

Conclusion

Multidisciplinary management between the anaesthetic, obstetric and neurosurgical teams should be established ideally before conception. Patients with a surgically corrected ACM should have a MRI postsurgery. If the MRI is not available, an effort to engage in a consultation with the neurosurgeon is still necessary even if they are asymptomatic. There should be an individualized approach to parturients with uncommon, non-pregnancy-specific disorders such as type 1 ACM. This case highlights that a spinal NA is a safe and effective option in parturients with a corrected ACM and syringomyelia undergoing a caesarean section, given that careful planning has been undertaken.

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Informed consent

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References

1. Bejjani GK. Definition of the adult Chiari malformation: a brief historical overview. *Neurosurg Focus* 2001; 11: E1
2. Snyder P. Chiari malformation and syringomyelia. *Radiol Technol* 2008; 79: 555–558.
3. Williams B. Malformations. In: Swash M and Oxybury J (eds). *Clinical neurology*. Edinburgh: Churchill Livingstone, 1991, pp. 1533–1582.
4. Chantigian RC, Koehn MA, Ramin KD, et al. Chiari I malformation in parturients. *J Clin Anesth* 2002; 14: 201–205.
5. Banerji NK and Millar JH. Chiari malformation presenting in adult life. Its relationship to syringomyelia. *Brain* 1974; 97: 157–168.
6. Paul KS, Lye RH, Strang FA, et al. Arnold-Chiari malformation. Review of 71 cases. *J Neurosurg* 1983; 58: 183–187.
7. Williams B. On the pathogenesis of syringomyelia: a review. *J R Soc Med* 1980; 73: 798–806.
8. Muir HA. General anaesthesia for obstetrics, is it obsolete? *Can J Anaesth* 1994; 41: R20–R25.
9. Barton JJ and Sharpe JA. Oscillopsia and horizontal nystagmus with accelerating slow phases following lumbar puncture in the Arnold-Chiari malformation. *Ann Neurol* 1993; 33: 418–421.
10. Hullander RM, Bogard TD, Leivers D, et al. Chiari I malformation presenting as recurrent spinal headache. *Anesth Analg* 1992; 75: 1025–1026.
11. Landau R, Giraud R, Delrue V, et al. Spinal anesthesia for cesarean delivery in a woman with a surgically corrected type I Arnold Chiari malformation. *Anesth Analg* 2003; 97: 253–255.
12. Mueller DM and Oro J. Chiari I malformation with or without syringomyelia and pregnancy: case studies and review of the literature. *Am J Perinatol* 2005; 22: 67–70.
13. Hilt H, Gramm H-J and Link J. Changes in intracranial pressure associated with extradural anaesthesia. *Br J Anaesth* 1986; 58: 676–680.
14. Nogues M, Delorme R, Saadia D, et al. Postural tachycardia syndrome in syringomyelia: response to fludrocortisone and beta-blockers. *Clin Auton Res* 2001; 11(4): 265–267.
15. Agusti M, Adalia R, Fernandez C, et al. Anaesthesia for caesarean section in a patient with syringomyelia and Arnold-Chiari type I malformation. *Int J Obstet Anesth* 2004; 13: 114–116.