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CASE REPORTS/CASE SERIES

Epidural anesthesia for Cesarean delivery in a patient with post-traumatic cervical syringomyelia

Anesthésie péridurale pour un accouchement par césarienne chez une patiente atteinte d'une syringomyélie cervicale post-traumatique

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Abstract

Purpose *To illustrate the successful management of a patient with post-traumatic syringomyelia (PTS) and chronic pain syndrome who presented for elective Cesarean delivery under epidural anesthesia.*

Clinical findings *A 30-yr-old gravida 3 para 1 woman, with a known diagnosis of cervical PTS secondary to a whiplash injury sustained three years earlier, presented to the labour and delivery unit at 31 weeks' gestation. She had severe pain in the cervical and lumbar spine, motor and sensory deficits in the upper extremities, tender mass in her left trapezius muscle, and history of dizziness and syncopal episodes. She was taking oxycodone 120 mg·day^{-1} Magnetic resonance imaging of her spine revealed a syrinx of 2 mm in diameter extending from C4 to T1 levels with disc protrusions in the C4-C6 region. There was no evidence of Arnold-Chiari malformation or elevated intracranial pressure. On airway examination, her Mallampati score appeared normal, but there was a limitation in the range of her neck movements in all directions. An elective Cesarean delivery was planned at 39 weeks' gestation. An epidural catheter was placed using ultrasound guidance, and the procedure was performed without complications.*

Conclusions *The successful management of this case suggests that epidural can be considered in women with cervical PTS presenting for a Cesarean delivery.*

Résumé

Objectif *Présenter la prise en charge réussie d'une patiente atteinte de syringomyélie post-traumatique (SPT) et d'un syndrome de douleur chronique lors d'un accouchement par césarienne non urgent sous anesthésie péridurale.*

Constatations cliniques *Une femme de 30 ans gravida 3 para 1, chez qui un diagnostic préalable de SPT avait été posé à la suite d'un coup de fouet cervical trois ans plus tôt, s'est présentée à l'unité de travail obstétrical et d'accouchement à 31 semaines de grossesse. Elle souffrait de douleurs graves au niveau de la colonne cervicale et de la colonne lombaire et de troubles moteurs et sensoriels dans les membres supérieurs. En outre, elle présentait une masse douloureuse dans son trapèze gauche et avait des antécédents de vertiges et de syncopes. Elle prenait de l'oxycodone 120 mg·jour^{-1} L'imagerie par résonance magnétique de sa colonne a révélé un syrinx de 2 mm de diamètre allant des vertèbres C4 à T1 ainsi que d'une protrusion des disques dans la région C4-C6. Aucun signe de malformation d'Arnold-Chiari ou de pression intracrânienne élevée n'a été observé. À l'examen des voies aériennes, son score de Mallampati semblait normal, mais l'ampleur de ses mouvements du cou était limitée dans toutes les directions. Un accouchement par césarienne non urgent a été planifié à 39 semaines de grossesse. Un cathéter péridural a été placé par échoguidage, et l'intervention a été réalisée sans complications.*

Conclusion *La prise en charge réussie de ce cas indique que l'anesthésie péridurale peut être envisagée chez les*

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femmes atteintes de SPT cervicale lors d'un accouchement par césarienne.

Syringomyelia is the progressive formation of a cavity or syrinx within the spinal cord that compresses the grey spinal cord tissue causing a variety of neurological symptoms.[1] It has traditionally been categorized as either congenital or acquired. The congenital form is associated with Arnold-Chiari malformation and is the most common presentation (80-85%).[2] This is referred to as "communicating" syringomyelia due to an initial or persisting continuity between the syrinx and cerebrospinal fluid (CSF) in the central canal of the spinal cord. The acquired form, also known as "non-communicating" syringomyelia, can be related to many factors, including inflammatory, neoplastic, and traumatic etiologies.

Post-traumatic syringomyelia (PTS) is typically seen following trauma to the spinal cord, and many theories have been proposed to explain its pathogenesis,[3,4] including cavitation following spine ischemia or infarction, arachnoiditis, inflammatory response, and intraparenchymal hematoma and its subsequent absorption leaving a cystic cavity.[5,6] According to Di Lorenzo *et al.*,[2] all types of syringomyelia have a common underlying cause, i.e. the alteration of normal CSF flow due to blockage at some level of the subarachnoid space.

The prevalence of syringomyelia was estimated to be around 9 per 100,000, and 25% of those were associated with PTS.[5] It is important to note that this estimation was proposed prior to the widespread use of magnetic resonance imaging (MRI), so the actual prevalence is likely to be much higher. A prevalence as high as 130 per 100,000 has been reported recently in some regions of the Russian Federation.[7,8] It is estimated that 1-9% of patients with spinal cord trauma have the likelihood of developing symptomatic syringomyelia within 30 years of trauma.[5]

Although a few cases of peripartum management of patients with syringomyelia have been reported in the literature, these reports are described in the context of congenital form and not PTS.[1,9-14] We present the successful peripartum anesthetic management of a parturient suffering from PTS. The patient gave written informed consent for the publication of this case report.

Case presentation

A 30-yr-old woman, gravida 3 para 1, presented to the preadmission clinic at Mount Sinai Hospital for anesthetic consultation at 31 weeks' gestation. She was diagnosed with cervical PTS secondary to a whiplash injury to her spine sustained in a motor vehicle accident three years earlier.

Her symptoms started within a few days after the accident and involved pain in the cervical spine and torticollis toward the right side. The pain in her cervical spine evolved over time to include upper and lower thoracic as well as lumbar spine and paraspinal areas. Her back pain increased significantly with Valsalva maneuvers, such as bearing down, coughing or sneezing, and prevented her from walking normally or sitting upright in a chair. She had decreased sensations below C2 dermatome and diffuse non-focal weakness involving her arms, mainly the left. She also experienced a few spontaneously resolving syncopal episodes which were preceded by severe pain and dizziness. She denied other symptoms of autonomic dysfunction.

Eight months following her accident, she was referred to a neurosurgeon for further investigations. Magnetic resonance imaging of her spine revealed a cervico-thoracic syrinx of 2 mm in diameter extending from C4 to T1 level with disc protrusions in the C4-C6 region indenting the thecal sac without cord compression (Figure). There was no evidence of Arnold-Chiari malformation or raised intracranial pressure (ICP).

The patient also had a history of childhood asthma, sickle cell trait, depression, and anxiety. Her medications were fluoxetine, trazadone, clonazepam, famotidine, and oxycodone 120 mg daily (40 mg in the morning and 80 mg at night). She smoked medicinal marijuana two to three times a day. On physical examination at 31 weeks' gestation, she was found to be 165 cm tall and her weight was 84.5 kg. She was distressed because of her back pain, and she walked slowly with some limp. Her vital signs were stable, and airway examination revealed Mallampati class 2 score with limitations in the range of her neck movements in all directions due to a firm mass in the left shoulder area and torticollis. Her spinous processes could not be palpated due to spasm of her paravertebral muscle columns. Her lumbar spine was lordotic, and she could not voluntarily reverse this lordosis because of her back pain. No evidence of scoliosis was found.

In preparation for her delivery, thorough planning was carried out by a team consisting of an obstetrician, an anesthesiologist, a neurosurgeon, a neonatologist, and a psychiatrist. Another MRI of her spine was ordered, which showed no changes in the size of her syrinx. After consultation with the neurosurgeon, obstetrician, and the patient, it was decided to perform an elective Cesarean delivery at $39^{3/7}$ weeks' gestation. We discussed the options of both general and regional anesthesia with the patient and informed her about the lack of anesthesia literature in patients with PTS. The risks of changes in CSF dynamics and pressure during tracheal intubation and spinal anesthesia were explained to the patient. The usual risks of general anesthesia in pregnant patients, such as

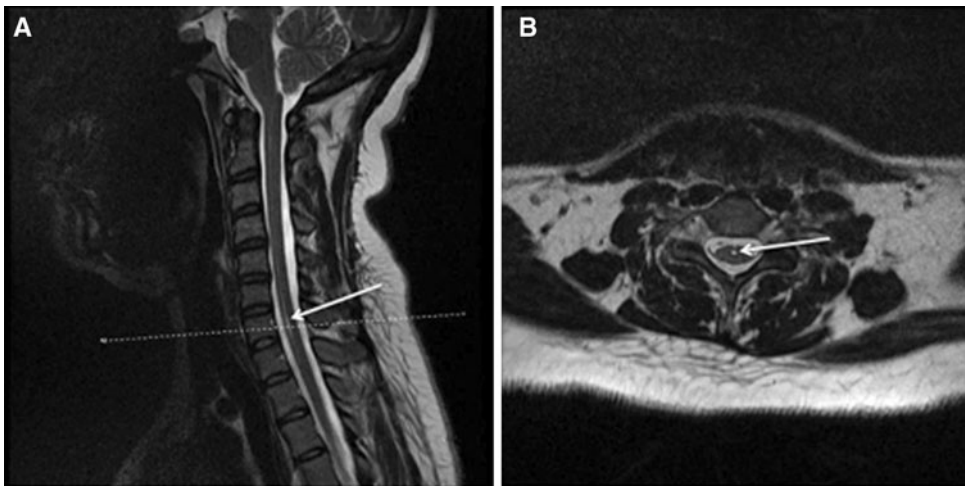

Figure Magnetic resonance imaging scan of the spinal cord at the cervicothoracic level revealing a 2-mm dilated syrinx in the central canal from C4 to T1 (arrow at C7 level). Sagittal view (A) and axial view (B). There is no Arnold-Chiari malformation

possible aspiration and difficult tracheal intubation were also discussed. Titrated epidural anesthesia was considered a preferred choice due to the possibility of less CSF pressure change and the ability to use the epidural catheter postoperatively for pain management. Both the patient and the obstetrician were informed about the risk of unintentional dural puncture and its unknown consequences in patients with PTS. We reassured both of them that measures would be taken to minimize these risks, and we agreed to have the most senior anesthesiologist available perform the epidural procedure.

There was no change in the patient's neurological condition or vital signs on the day of the planned Cesarean delivery. Premedication of ranitidine 50 mg and metoclopramide 10 mg was given intravenously. Epidural anesthesia was performed with the patient in the sitting position after determining the L3-4 interspace, the needle insertion point, and the estimated depth to the epidural space by ultrasonography. To position her comfortably, she was administered fentanyl 100 μg intravenously in aliquots of 50 μg each. Using an 18G Tuohy needle, the epidural space was located through the pre-determined insertion point by loss of resistance to saline technique. Anesthesia was established by slowly injecting 2% lidocaine with epinephrine 1:200,000 (in aliquots of 3-5 mL for a total of 25 mL) with fentanyl 50 μg. A bilateral sensory block height of T4 was achieved. The patient's preoperative blood pressure was 120/60 mmHg and her heart rate was 85 beats·min^{-1}. Her blood pressure was maintained within 10% of the baseline levels throughout the procedure using vasopressors (intravenous phenylephrine 400 μg total) and lactated Ringer's solution (1,500 mL total). A healthy neonate was delivered with Apgar scores of 8 and 9 at one and five minutes, respectively; the neonate did not require ventilatory assistance. After the delivery, the patient received an intravenous infusion of oxytocin (20 units·L^{-1} at the rate of 150 mL hr^{-1}), epidural morphine 2.5 mg, ketorolac 30 mg *iv*, and acetaminophen 1,300 mg rectally. She was given oral diclofenac 50 mg every eight hours and acetaminophen 1 g every six hours in addition to her daily analgesic medications. Patient-controlled analgesia was offered, but the patient refused it. She received a total of 20 mg of intravenous morphine for breakthrough pain during the first two days postpartum. Her abdominal pain was under control; however, she continued to have back pain. No other complications were encountered, and she was discharged three days after the Cesarean delivery.

Discussion

Our patient presented with the diagnosis of PTS with chronic pain syndrome and was managed successfully for Cesarean delivery under epidural anesthesia.

Patients with syringomyelia may present with diverse pathophysiology and clinical features. The clinical manifestations include altered pain and temperature sensations in the upper limbs (lateral spinothalamic tracts), flaccid weakness in the upper limbs and shoulder girdle (anterior horn cells), and spastic paresis in the lower limbs (corticospinal tracts). Weakness of the paraspinal musculature may lead to thoracic scoliosis.[2] The presence of posterior column signs usually signifies advanced disease. Involvement of the autonomic nervous system is not uncommon. Syringomyelia associated with Arnold-Chiari malformation may present with signs of increased ICP. As these two conditions are closely associated with each other in the majority of cases, Arnold-Chiari malformation must be ruled out when syringomyelia is diagnosed.[2] These patients should undergo a comprehensive preoperative assessment that includes the degree of neurological deficits and, signs of increased ICP and autonomic neuropathy.

The CSF flow dynamics in patients with syringomyelia may have implications during labour, delivery, and

anesthetic management of these patients. The pathophysiology of the development of a syrinx is still poorly understood. An "intramedullary pulse pressure theory" has been proposed recently to explain the hydrodynamics of syringomyelia. The theory suggests that a syrinx is caused by increased pulse pressure in the spinal cord that is greater than that in the nearby subarachnoid space.[15] The pressure in the syrinx is higher than that in the surrounding CSF and is influenced by changes in subarachnoid space compliance. In patients with PTS, the presence of a locally tethered cord or a blockage by arachnoid scarring may diminish subarachnoid space compliance, which can lead to an increase in CSF inflow. Consequently, syrinx enlargement, cord enlargement above or below obstruction, or cord dissection can occur.[5,15]

It is possible that the process of labour itself, similar to coughing and straining, can be a precipitating factor for syrinx enlargement.[16] Therefore, in our case, Cesarean delivery was preferred over vaginal delivery to avoid sudden clinical deterioration and worsening of neurological symptoms. Although vaginal delivery is not contraindicated, an assisted second stage is advocated.[17]

The challenges in the anesthetic management of our case included: (a) potential difficulty in the placement of regional block due to lumbar spine hyperlordosis, pain, and paraspinal muscle spasms; (b) considerations for avoiding pressure changes within the syrinx due to administration of spinal or epidural drugs and pressor response during general anesthesia; (c) potential difficulty in tracheal intubation due to a dystonic neck mass; and (d) difficult postoperative pain management due to the patient's chronic high-dose opioid use.

Spinal anesthesia may not be a preferred option for Cesarean delivery in patients with PTS, as abrupt changes in CSF pressure as well as severe hypotension are to be avoided, especially in the event of disautonomia. Spinal anesthesia could also produce unpredictable effects due to abnormal CSF flow dynamics in these patients. Two case reports of aggravated signs and symptoms within two weeks of dural puncture have been described in syringomyelia associated with Arnold-Chiari malformation.[18-20]

Daskalakis *et al.*[1] described an uncomplicated elective Cesarean delivery under general anesthesia in the presence of PTS. The authors discussed both general and epidural anesthesia as possible techniques. However, several critical issues have to be considered while providing general anesthesia in such patients, including avoidance of increase in ICP and pressure changes in the syrinx as a result of pressor response during intubation, coughing, vomiting, hypoxia, or hypercarbia. Close hemodynamic monitoring is also necessary in patients with autonomic neuropathy. Other issues with the administration of general anesthesia include ventilation-perfusion abnormalities due to vertebral column deformities and an unpredictable response to muscle relaxants in patients with neurological involvement.[21,22] Our patient had severe torticollis and a dystonic muscle mass in the neck, which might have posed an additional problem during airway intervention.

Epidural anesthesia was chosen in our case as it may offer several advantages over spinal and general anesthesia in patients with PTS. These benefits include avoidance of the potential hazards of securing the airway, reduced incidence of hypotension and deterioration of autonomic neuropathy, and minimal change in the existing CSF pressure relationship if the medication is titrated gradually. The difficulty in epidural placement due to the hyperlordotic state in our patient was overcome by prior administration of intravenous fentanyl and use of ultrasound for accurate localization of the landmarks. Moreover, the procedure was performed carefully by a senior anesthesiologist and not by a trainee. Although the possibility of accidental dural puncture with an epidural needle and subsequent alterations in CSF pressures cannot be ruled out, the above measures decreased the likehood of such events. There is still a possibility of sudden distension of the epidural space due to the local anesthetic injection, which in turn could induce subarachnoid space compression thereby increasing ICP and creating a potentially damaging pressure wave within the syrinx.[10,23,24] To minimize this problem, gradual titration of local anesthetic was instituted through an epidural catheter. Considering the patient's chronic pain syndrome, the added advantage of placing an epidural catheter was to provide additional local anesthetics and opioids if required in the postoperative period.

The successful management of this case suggests that an epidural can be considered in women with post-traumatic cervical syringomyelia presenting for a Cesarean delivery.

Conflict of interests None declared.

References

1. *Daskalakis GJ*, *Katsetos CN*, *Papageorgiou IS*, *et al*. Syringomyelia and pregnancy-case report. Eur J Obstet Gynecol Reprod Biol 2001; 97: 98-100.
2. *Di Lorenzo N*, *Cacciola F*. Adult syringomielia. Classification, pathogenesis and therapeutic approaches. J Neurosurg Sci 2005; 49: 65-72.
3. *Carroll AM*, *Brackenridge P*. Post-traumatic syringomyelia: a review of the cases presenting in a regional spinal injuries unit in the north east of England over a 5-year period. Spine (Phila Pa 1976) 2005; 30: 1206-10.
4. *Williams B*, *Sgouros S*, *Nenji E*. Cerebrospinal fluid drainage for syringomyelia. Eur J Pediatr Surg 1995; 5: 27-30.
5. *Brodbelt AR*, *Stoodley MA*. Post-traumatic syringomyelia: a review. J Clin Neurosci 2003; 10: 401-8.

6. *Wozniewicz B*, *Filipowicz K*, *Swiderska SK*, *Deraka K*. Pathophysiological mechanism of traumatic cavitation of the spinal cord. Paraplegia 1983; 21: 312-7.
7. *Bogdanov EI*, *Heiss JD*, *Mendelevich EG*. The post-syrinx syndrome: stable central myelopathy and collapsed or absent syrinx. J Neurol 2006; 253: 707-13.
8. *Bogdanov EI*, *Mendelevich EG*. Syrinx size and duration of symptoms predict the pace of progressive myelopathy: retrospective analysis of 103 unoperated cases with craniocervical junction malformations and syringomyelia. Clin Neurol Neurosurg 2002; 104: 90-7.
9. *Roelofse JA*, *Shipton EA*, *Nell AC*. Anaesthesia for caesarean section in a patient with syringomyelia. A case report. S Afr Med J 1984; 65: 736-7.
10. *Nel MR*, *Robson V*, *Robinson PN*. Extradural anaesthesia for caesarean section in a patient with syringomyelia and Chiari type I anomaly. Br J Anaesth 1998; 80: 512-5.
11. *Lopez R*, *Nazar C*, *Sandoval P*, *Guerrero I*, *Mellado P*, *Lacassie HJ*. Neuraxial analgesia during labor in a patient with Arnold-Chiari type I malformation and syringomyelia (Spanish). Rev Esp Anestesiol Reanim 2007; 54: 317-21.
12. *Chantigian RC*, *Koehn MA*, *Ramin KD*, *Warner MA*. Chiari I malformation in parturients. J Clin Anesth 2002; 14: 201-5.
13. *Mueller DM*, *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.
14. *Murayama K*, *Mamiya K*, *Nozaki K*, *et al*. Cesarean section in a patient with syringomyelia. Can J Anaesth 2001; 48: 474-7.
15. *Greitz D*. Unraveling the riddle of syringomyelia. Neurosurg Rev 2006; 29: 251-63.
16. *Pearce JM*. Surgical management of syringomyelia. Br Med J (Clin Res Ed) 1981; 283: 1204-5.
17. *Parker JD*, *Broberg JC*, *Napolitano PG*. Maternal Arnold-Chiari type I malformation and syringomyelia: a labor management dilemma. Am J Perinatol 2002; 19: 445-50.
18. *Hullander RM*, *Bogard TD*, *Leivers D*, *Moran D*, *Dewan DM*. Chiari I malformation presenting as recurrent spinal headache. Anesth Analg 1992; 75: 1025-6.
19. *Barton JJ*, *Sharpe JA*. Oscillopsia and horizontal nystagmus with accelerating slow phases following lumbar puncture in the Arnold-Chiari malformation. Ann Neurol 1993; 33: 418-21.
20. *Preston R*. Spinal cord disorders. *In*: Gambling DR (Ed.). Obstetric Anesthesia and Uncommon Disorders. 2nd ed. Cambridge: Cambridge University Press; 2008.
21. *Agusti M*, *Adalia R*, *Fernandez C*, *Gomar C*. Anaesthesia for caesarean section in a patient with syringomyelia and Arnold-Chiari type I malformation. Int J Obstet Anesth 2004; 13: 114-6.
22. *Fiacchino F*, *Gemma M*, *Bricchi M*, *Giombini S*, *Regi B*. Sensitivity to curare in patients with upper and lower motor neurone dysfunction. Anaesthesia 1991; 46: 980-2.
23. *Williams B*. On the pathogenesis of syringomyelia: a review. J R Soc Med 1980; 73: 798-806.
24. *Hilt H*, *Gramm HJ*, *Link J*. Changes in intracranial pressure associated with extradural anaesthesia. Br J Anaesth 1986; 58: 676-80.