Case Report

Anesthetic Management of a Patient with Chiari Malformation Type I and Syringomyelia Undergoing Total Hip Replacement

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Abstract

Syringomyelia associated with Chiari malformation type I presents significant anesthetic challenges due to risks related to altered cerebrospinal fluid dynamics, autonomic dysfunction, impaired respiratory function, and potential for increased intracranial pressure. Evidence-based recommendations remain limited, particularly for non-obstetric procedures. We report the successful anesthetic management of a 52-year-old male with Chiari malformation type I and cervicothoracic syringomyelia undergoing elective total hip arthroplasty. Given prior posterior fossa decompression, thoracic kyphoscoliosis, and concern for increased intracranial pressure during airway manipulation, spinal anesthesia was selected. The procedure and postoperative recovery were uneventful, with stable hemodynamics and no neurological deterioration. This case supports the growing view that neuraxial anesthesia can be safely performed in selected patients with syringomyelia and Chiari malformation type I when there is no radiological evidence of elevated intracranial pressure or syrinx extension at the puncture site. Individualized anesthetic planning, multidisciplinary collaboration, and meticulous intraoperative monitoring are key to optimizing outcomes in non-obstetric surgery.

Keywords: Arnold-Chiari malformation; syringomyelia; spinal anesthesia; total hip replacement; individualized anesthetic management

1 Introduction

Syringomyelia is a neurological disorder characterized by the presence of a fluid-filled cyst, or syrinx, within the spinal cord. The most common location of syrinx formation is between C2 and Th9, although it can occur at any level. Patients may be asymptomatic or may present with pain, scoliosis, neurological deficits, or autonomic dysfunction. Symptoms depend on the size and location of the syrinx (1). There are two types of syringomyelia: congenital and acquired.

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Congenital syringomyelia is frequently associated with Chiari malformation (2). Acquired syringomyelia may result from spinal cord tumors, inflammation (e.g., transverse myelitis or multiple sclerosis), infection (e.g., meningitis or arachnoiditis), or spinal cord injury.

Chiari malformations are structural abnormalities of the craniocervical junction, characterized by downward displacement of the lower part of the brain. The most common type is Chiari malformation type I, which is defined as herniation of the cerebellar tonsils through the foramen magnum (1,2). Posterior fossa hypoplasia and impaired cerebrospinal fluid (CSF) flow contribute to syrinx development in up to 25–75% of patients (1,3). Myelomeningocele and childhood-onset hydrocephalus are commonly associated with Chiari type II malformation (2). Syringomyelia associated with Chiari malformation may present with a broad spectrum of clinical manifestations, including neurological symptoms, features of autonomic dysfunction, scoliosis, and signs of increased intracranial pressure (4,5,6,7,8,9).

According to Costa et al., conservative management is recommended for patients who are asymptomatic or have mild symptoms and show radiologic confirmation on MRI (1,2). Indication for surgical treatment of Arnold Chiari malformation include myelopathy, upper cervical cord symptoms and brainstem symptoms (10). Surgical treatment focuses on correcting the malformation and restoring cerebrospinal fluid flow at the foramen magnum, with the choice of technique determined by the mechanism of hindbrain herniation (10).

Although recommendations for the anesthetic management of these patients exist, official and comprehensive guidelines are still lacking. The anesthetic management of such patients remains challenging, as both general and neuraxial anesthesia carry potential risks. General anesthesia may be complicated by difficult airway management, increased intracranial pressure, autonomic dysfunction, and ventilatory impairment due to scoliosis. Neuraxial anesthesia, on the other hand, raises concerns regarding alterations in cerebrospinal fluid dynamics and technical difficulties related to spinal deformities. Here, we present the anesthetic management of a patient with Chiari malformation and syringomyelia who underwent hip replacement surgery.

2 Case Report

A 52-year-old male with left-sided coxarthrosis, Chiari malformation type I and syringomyelia was scheduled for total hip replacement surgery. His comorbidities included hypopituitarism, hyperprolactinemia, osteopenia, thoracic kyphoscoliosis, and chronic lumbosacral syndrome. His regular medications were hydrocortisone, levothyroxine sodium, cholecalciferol, naproxen sodium, and testosterone decanoate every three months.

Eight years earlier, he had presented with right-sided neck pain radiating to the shoulder and arm, followed by progressive weakness of the right arm and leg. MRI of the brain and cervical spine showed a centrally located syrinx throughout the cervical spinal cord and features consistent with Arnold–Chiari malformation. Thoracic MRI revealed syringohydromyelia extending from the cervical cord to Th7, while lumbar MRI showed a normal spinal canal. He underwent suboccipital craniectomy and C1 decompressive laminectomy with duraplasty under general

anesthesia. Postoperatively, there was no progression of neurological symptoms, and a follow-up MRI two years later showed no change.

At presentation for the current surgery, the patient reported severe left hip pain and quadriparesis. Preoperative chest X-ray revealed thoracic scoliosis; ECG and laboratory tests were within normal limits. Airway assessment showed Mallampati grade II with adequate neck mobility. Premedication included oral midazolam 7.5 mg, hydrocortisone 15 mg, levothyroxine sodium 100 μg, pantoprazole 40 mg, and intravenous sodium hydrocortisone succinate 50 mg.

The surgery was performed under spinal anesthesia at L4–L5 using a 25-G Quincke needle and 0.5% bupivacaine. Sedation was maintained with intermittent boluses of midazolam. Intraoperative monitoring included non-invasive blood pressure, ECG, and SpO_2. The surgical procedure lasted 2 hours. During the surgical procedure, the patient remained hemodynamically stable, with an average blood pressure of 125/90 mmHg and a heart rate of approximately 80/min. The patient received a total of 1500 ml of crystalloids. At the end of surgery, intravenous sufentanil 5 μg was administered.

In the recovery room, an ultrasound-guided fascia iliaca block was performed using 30 mL of 0.25% levobupivacaine. Postoperative intravenous analgesia consisted of paracetamol 1 g three times daily, ketoprofen 100 mg twice daily, and tramadol 100 mg three times daily. The patient remained hemodynamically stable, pain-free, and with sufficient respiration, and was subsequently transferred to the surgical ward.

3 Discussion

The choice of anesthesia for patients with syringomyelia, particularly when associated with Chiari malformation type I (CM-I), remains clinically challenging due to variable neurological presentations and the absence of universal guidelines. Optimal anesthetic planning relies on detailed preoperative assessment including neurological status, autonomic involvement, airway anatomy, respiratory function and MRI findings (3,11,12). With the increasing use of MRI, more asymptomatic cases of syringomyelia are being identified, yet symptomatic patients commonly present with dissociated anesthesia, upper-limb weakness, and spasticity of the lower extremities when the syrinx extends caudally (4). When syringomyelia coexists with Chiari malformation type I, patients may also present with symptoms of raised intracranial pressure (ICP), such as headache, vomiting, dizziness, dysphagia, altered mental status, visual changes, hypertension, bradycardia, and irregular respiration (4,9).

Autonomic dysfunction—including bowel and bladder disturbances, impaired circulatory reflexes, postural hypotension, bradycardia, and tachycardia—may significantly increase the risk of intraoperative hemodynamic instability (5,6,7,8). Many authors emphasize the importance of early recognition of autonomic disturbances, as it provides awareness of the need for intraoperative invasive monitoring and thus enables timely identification of hemodynamic instability, as well as preparedness for sudden events (3,8). Chronic pain in the neck, shoulders, and arms is frequent, and scoliosis may develop, particularly in pediatric patients (4).

Both general anesthesia and neuraxial anesthesia carry specific advantages and disadvantages.

General anesthesia may be complicated by reduced cervical mobility, difficult airway management and impaired ventilation in the presence of scoliosis. The videolaryngoscopy or fiberoscopy is often recommended to minimize head/neck manipulation (13,14). Intubation and coughing may transiently increase ICP. Abnormal responses to neuromuscular blocking agents and blunted cardiovascular responses to hypovolemia further necessitate vigilant hemodynamic monitoring and careful anesthetic titration (3,15,16).

Neuraxial anesthesia has its own challenges. Anatomical deformities such as scoliosis and paravertebral muscle spasm can make spinal or epidural puncture technically difficult (4). Concerns have been raised about altering CSF flow dynamics and syrinx pressure following administration of local anesthetics, particularly rapid bolus injections (3,4,13). There is a theoretical risk of neurological deterioration resulting from cerebrospinal fluid shifts or increased pressure within the syrinx cavity. As with general anesthesia, hemodynamic monitoring is crucial because of the risk of hypotension. Evidence from obstetric cases suggests that neuraxial anesthesia can be safe in carefully selected patients—even after posterior fossa decompression—provided multidisciplinary evaluation and imaging review are performed (3,12,17,18).

Our non-obstetric case contrasts with prior reports that predominantly involve parturients. Most published case reports and reviews concerning anesthesia in patients with syringomyelia and Chiari malformation involve obstetric procedures. For many years, pregnant women with syringomyelia and Chiari malformation have been managed successfully. Teo et al. described a parturient with residual syringomyelia after surgical decompression who successfully underwent elective cesarean section under spinal anesthesia. The authors favored a fine-gauge spinal needle and avoided large epidural boluses (which may transiently increase ICP), recommending slow, incremental epidural dosing when an epidural is chosen (19). The paper highlights the importance of a multidisciplinary approach (anesthesiologist, obstetrician, neurosurgeon) and careful monitoring of patients with residual neurological symptoms. The authors conclude that spinal anesthesia can be safely performed in selected cases, although consensus and guidelines are still lacking (19).

Waters et al. reported that the majority of patients received neuraxial anesthesia without any neurological complications or signs of increased intracranial pressure. These findings suggest that, in the absence of elevated intracranial pressure, neuraxial techniques, including epidural and spinal anesthesia, can be considered safe and appropriate (12). Several other studies have suggested that vaginal delivery and neuraxial anesthesia administration can be safely performed in women with Chiari I malformation (17,18). Sastry et al. recommended consideration of decompression before pregnancy for patients with significant neurologic symptoms and for asymptomatic patients recommended vaginal delivery under neuraxial anesthesia (11). They also recommended minimizing maternal Valsalva efforts, either through cesarean delivery under general or neuraxial anesthesia, or by performing assisted vaginal delivery under neuraxial anesthesia (11).

Regarding non-obstetric cases, there are case reports available, but no systematic reviews.

For example, Anand et al. described pronounced hemodynamic instability (profound hypotension with compensatory tachycardia) precipitated by prone positioning in a patient with Chiari I malformation and syringomyelia undergoing foramen magnum decompression—managed with fluids and phenylephrine infusion—whereas our patient remained stable throughout hip arthroplasty in the lateral decubitus position under spinal anesthesia (8). Nevertheless, caution is warranted when performing neuraxial anesthesia in patients with underlying but undiagnosed syringomyelia and Chiari malformation, as neurological deterioration may manifest only postoperatively.

Madhaw et al. reported a patient who developed paraparesis due to dorsal spinal cord involvement and syrinx formation following spinal anesthesia for abdominal surgery, emphasizing that although lumbar spinal or epidural techniques generally carry a lower complication rate compared to general anesthesia, the risk of spinal cord injury and subsequent neurological sequelae, though rare, remains a concern (20). Because syringomyelia most commonly involves the cervical spinal cord and less frequently the thoracic region, spinal anesthesia is generally considered safe, as it is routinely performed at the lumbar level where syrinx formation is rare. However, in cases where thoracic epidural anesthesia is contemplated, pre-procedural MRI should be performed to accurately determine the localization of the cysts and avoid potential neurological complications.

Preoperative evaluation should include assessment of neurological status, autonomic dysfunction, respiratory function, and MRI findings, and should be conducted by a multidisciplinary team comprising an anesthesiologist, a neurologist, and a neurosurgeon (3,21). Clinical consequences of anesthetic management in patients with syringomyelia include hemodynamic instability, such as hypotension, tachycardia, and bradycardia, as well as the potential risk of worsening pre-existing neurological deficits. These patients are also particularly vulnerable to complications related to airway manipulation and intraoperative positioning, especially when placed in the prone position.

Current literature strongly reinforces individualized decision-making, emphasizing multidisciplinary planning and involvement of experienced anesthesiologists, particularly in cases with craniovertebral abnormalities raising concerns about airway complexity or autonomic instability (22,23). Video- or fiberoptic-assisted airway management, avoidance of abrupt increases in ICP, and invasive blood pressure monitoring are recommended strategies in neurosurgical and non-obstetric settings (24,25). Advanced imaging, including CSF flow studies, may further support risk stratification in patients with persistent tonsillar descent or progressive syrinx expansion (22). Given these challenges, there are no universal guidelines for anesthetic management in this population. Instead, the approach should be individualized, based on a thorough preoperative evaluation, multidisciplinary planning, and meticulous intra- and postoperative monitoring.

Orthopedic procedures in patients with syringomyelia and CM-I pose unique challenges. Thoracic deformity and scoliosis can compromise respiratory mechanics and predispose to hypoventilation and postoperative pulmonary complications (21,25). Lateral or prone positioning must be optimized to avoid worsening spinal cord compression, with careful protection of the cervical spine (24). Chronic syrinx-related weakness and reduced bone density may increase fracture risk and influence postoperative mobility (22). Neuraxial anesthesia may be advantageous

in orthopedic surgery by reducing airway manipulation and preserving respiratory function, provided slow incremental dosing is used to prevent adverse CSF pressure shifts (22,23).

In this patient, we chose spinal anesthesia because of several considerations: (1) better hemodynamic stability, (2) prior decompression surgery and potential airway difficulties, (3) risk of increased ICP during intubation, (4) thoracic scoliosis and potential respiratory dysfunction and ventilation problems, and (5) syrinx localization limited to the cervical and thoracic cord.

Historically, general anesthesia was favored, but neuraxial techniques are increasingly reported as safe. In available literature, spinal or epidural anesthesia has not been associated with adverse outcomes or worsening of syringomyelia (13,19,26). Notably, most published reports on anesthesia in syringomyelia involve obstetric patients; non-obstetric case descriptions—such as the present hip arthroplasty—are comparatively scarce in the literature.

4 Conclusion

In conclusion, contemporary evidence indicates that neuraxial anesthesia can be safely performed in most patients with syringomyelia and CM-I when detailed neurological assessment, MRI review, and evaluation for ICP elevation are undertaken. Multidisciplinary planning, enhanced intraoperative monitoring, careful patient positioning, and slow incremental dosing are key to mitigating perioperative risk. Although most published experience derives from obstetric populations, the present case contributes to the limited non-obstetric data demonstrating that spinal anesthesia can be a safe and effective alternative to general anesthesia in appropriately selected patients.

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Ethics Statement

Written informed consent was obtained from the patient.

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Conflict of Interest

The authors declare that they have no conflicts of interest.

Author Contributions

M.L. wrote the manuscript. **T.M.T.** conceived and designed the case report. **G.S.** collected the clinical data and patient history. **F.J.** reviewed the literature. All authors critically revised the manuscript for important intellectual content. All authors read and approved the final manuscript.

Data Availability

Data sharing is not applicable to this article as no datasets were generated or analyzed.

Informed Consent

Written informed consent was obtained from the patient for the publication of this case report.

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