Indian Journal of Clinical Anaesthesia 2024;11(4):593–595

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Indian Journal of Clinical Anaesthesia

Journal homepage: www.ijca.in

Case Report

Anaesthetic management of a child with syringomyelia undergoing foramen magnum decompression: A case report

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ARTICLE INFO

Article history:
Received 02-09-2024
Accepted 14-10-2024
Available online 07-11-2024

Keywords:
Syringomyelia
Arnold Chiari malformation
Syrinx

ABSTRACT

Syringomyelia is a neurological disorder where a cyst consisting of fluid is formed within the spinal cord which results in various neurological symptoms. Treatment could be either conservative in the absence of neurologic symptoms or definitive surgical treatment. The most important task for an anesthesiologist is to provide general anesthesia with the use of intraoperative neuromuscular monitoring (IONM) for better neurological outcomes during foramen magnum decompression. Here through the case report of a 16-year-old male child with syringomyelia posted for foramen magnum decompression, we tend to focus upon anesthetic management of syringomyelia and the use of IONM and its benefits over routine management.

1. Introduction

Syringomyelia is a neurological condition where a cyst filled with fluid forms within the spinal cord parenchyma or canal. The name Syringomyelia is derived from two Greek words “channel” and “marroin” by Ollivier d’ Angers in 1827. The prevalence of syringomyelia is 8.4 per 1,00,000 and it occurs in males more than females, usually in the age between 25 to 40 years.[1] In syringomyelia the cerebrospinal fluid builds up within the spinal cord which leads to the expansion of the central canal and formation of syrinx resulting in compression of the surrounding nerve tissue causing neurological manifestations depending upon the size and location of the syrinx. Syrinx is developed when the normal cerebrospinal fluid flow in the spinal cord and brain stem is altered.[2] Arnold Chiari malformation is frequently associated with syringomyelia and must be assessed. Syringomyelia must be managed based on the severity and progression of symptoms. Syringomyelia can be managed conservatively if symptoms do not progress or worsen. The primary aim of surgical management is to remove the syrinx and reduce further spinal cord damage. Foramen magnum decompression surgery is recommended for patients with symptomatic or progressive syringomyelia. There are primarily two types of surgical treatment. One is to restore the normal cerebrospinal fluid flow around the spinal cord and the other is draining the syrinx.[3,4] Here we are presenting anesthetic management of a case of syringomyelia planned for foramen magnum decompression surgery.

2. Case Presentation

A 16-year-old male child who weighed about 42 kg and height of 145 cm presented with complaints of weakness in the left upper and lower limb for 4 years. The weakness was insidious in onset and gradually progressed. Magnetic Resonance Imaging (MRI) of cervical spine with whole spine screening suggested fluid filled cystic cavity extending from C1 – D11 vertebra suggestive of syringomyelia (Figure 1). Mild crowding of the foramen magnum and scoliotic deformity of the dorsal spine with

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https://doi.org/10.18231/j.ijca.2024.105

convexity towards the right side were also seen with Cobb's angle of 47 degree. Chest x-ray posteroanterior view showed scoliosis with convexity to right and deviation of tracheobronchial tree (Figure 2). On examination, the patient had difficulty in walking. Assessing motor power on the left upper limb found to be 4/5 and the left lower limb 4/5, including a decrease in tone. Motor power on the right upper limb was 5/5 in the right lower limb is 5/5. No abnormal findings in other systemic examination. The child was posted for foramen magnum decompression under general anesthesia. The child was premedicated with Tablet Alprazolam 0.25mg and Injection Pantoprazole 40 mg. Informed consent from the parents was obtained before shifting the child to the operating room. After instituting standard monitors, left radial artery was cannulated after performing modified Allen test for invasive blood pressure monitoring. Child was induced with Injection Thiopentone sodium 120mg, Injection Fentanyl 80mcg.Succinylcholine 80mg was given as muscle relaxant and trachea was intubated orally with 6.5mm flexo-metallic cuffed endotracheal tube. Since muscle relaxants would interfere in the neuromuscular monitoring it was given only for intubation and no additional doses were given. Total Intravenous Anesthesia with target-controlled infusion (TCI) pump with Propofol infusion targeted to serum level of 50mcg/kg/min and Fentanyl infusion at 40mcg/hour was used to maintain anaesthesia. The Patient was ventilated in Volume control ventilation with tidal volume of 400 ml, at 14 breaths per minute with FiO_2 50% and PEEP 4cm H_2O. Child was turned to prone position with close hemodynamic monitoring due to risk of autonomic dysfunction in syringomyelia and pressure points were padded to avoid injury. IONM with MEP was initiated to avoid injury to vital structures during the foramen magnum decompression. There were no challenges encountered during IONM with regard to positioning or depth of anesthesia. Intraoperative period was uneventful as common features associated with syringomyelia such as intraoperative bradycardia, hypotension or autonomic disturbances were not encountered. Child was extubated on table and shifted to post anaesthetic care unit. Postoperative analgesia was maintained with injection fentanyl 20 mcg/hr. Postoperative period was uneventful with no neurological deterioration and child was discharged on 5^{th} postoperative day.

3. Discussion

Syringomyelia commonly presents with Chiari malformation – a condition where tonsillar herniation occludes the flow of cerebrospinal fluid.[5] Syringomyelia might also be due to injury, tumors to spinal cord and damage caused by inflammation or rarely idiopathic. The two main types of syringomyelia, the congenital syringomyelia or communicating syringomyelia where

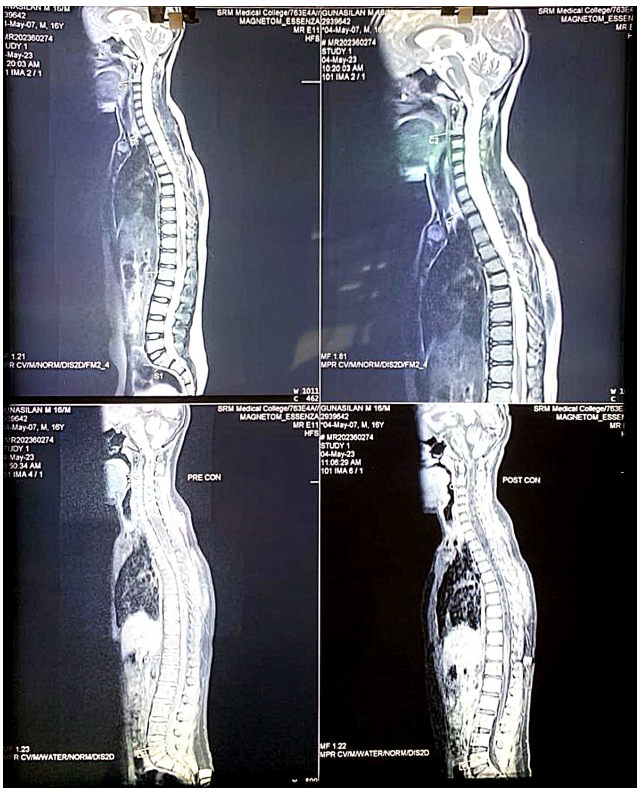

Figure 1: MRI cervical spine with whole spine screening showing a fluid filled cavity (Syrinx) from C1- D11 vertebral level MRI: Magnetic resonance imaging; C: Cervical; D: Dorsal

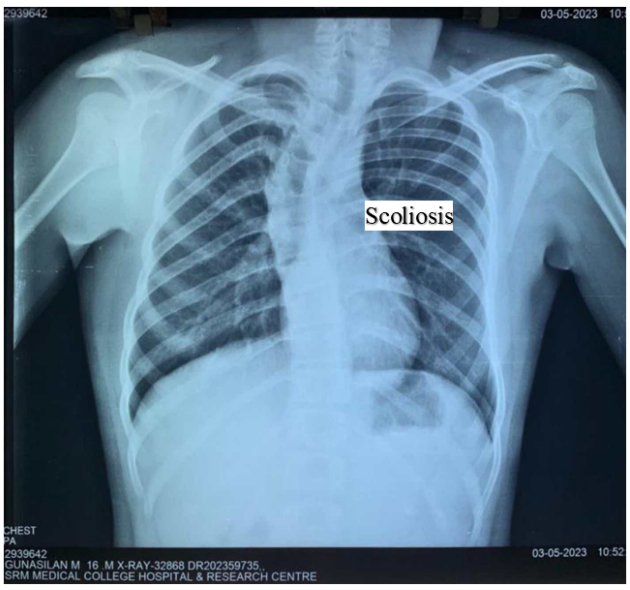

Figure 2: Chest X-ray PA view showing scoliosis PA: Posteroanterior

symptoms typically begin between 25 to 40 years and acquired type of syringomyelia which is otherwise called primary spinal syringomyelia or noncommunicating syringomyelia due to spinal cord injury, tumour, meningitis, arachnoiditis or tethered cord syndrome. The signs and symptoms depending on location, size and extent of syrinx are altered pain, temperature sensations in upper limbs (due to involvement of lateral spinothalamic tract), 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 muscles lead to thoracic scoliosis which could be the only symptom in children. Involvement of the autonomous nervous system is also common.

Intraoperative neurophysiological monitoring (IONM) is a tool to assess the function of neurological structures when surgery is being done consisting continuous monitoring of nerve tissue and localization of important neural structures. The aim of IONM is to find any neural damage occurring during surgery and to intervene quickly so as to decrease or to significantly reduce irreversible damage to the neural structures and thereby preventing permanent neurologic deficit. The use of neurophysiological monitoring during intraoperatively will require certain technique in anaesthesia to minimise interference and signal alteration caused by anaesthetic drugs. Different types of IONM are available that monitors a specific neural pathway, which includes somatosensory evoked potential (SSEP), motor evoked potential (MEP), brainstem auditory evoked potential (BAEP), visual evoked potential (VEP), Motor evoked potentials (MEP) are electrical signals observed in muscle or peripheral nerves after cortical stimulation. MEP is an important tool to confirm functional integrity of motor pathways during neurosurgeries.[6] MEPs monitors corticospinal tract starting with a stimulus in the motor cortex, then travels along the corticospinal tract to nerve root and the peripheral nerves before it terminates. Generating evoked potential is obtained by stimulating motor cortex with needle inserted on the scalp, directly stimulating the brain or with magnetic energy. Signal is delivered as train of many electrical impulses which modify the voltage to get acceptable signal. These stimulus pass through motor pathway of the anterior spinal cord before entering the peripheral nervous system and receiving action potential in muscle. Signal is measured with surface electrodes and needles in or around the targeted muscle. Usually, muscles monitored are abductor pollicis brevis in upper limb and tibialis anterior and gastrocnemius in lower limb. Alteration in signal due to anaesthesia and other systemic changes is global which impacts MEPs in extremities. MEP deteriorations due to surgery is usually focal.[7,8]

4. Conclusion

Effective management in a young child with syringomyelia requires a multidisciplinary team including experienced neurosurgeons, anesthesiologists, pediatricians and a full-fledged hospital and operation theatre setting. Syringomyelia is a serious condition which leads to progressive neurological symptoms and affects day to day activities. Preoperative comprehensive assessment and perioperative continuous monitoring may provide a successful outcome. Early detection and treating it conservatively or by surgery may provide a complete recovery. In addition to the routine procedure the use of IONM motor evoked potential MEP provides a safer procedure, and injury to nerve can be identified on table which otherwise could lead to catastrophic events postoperatively. Using IONM in future even for routinely done surgeries might bring an excellent outcome to the patient.

5. Source of Funding

None.

6. Conflict of Interest

None.

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Cite this article: Siddartha T, Kuppusamy A, Kumaran D. Anaesthetic management of a child with syringomyelia undergoing foramen magnum decompression: A case report. *Indian J Clin Anaesth* 2024;11(4):593-595.