CASE REPORT

Anesthetic management of a patient with syringomyelia and Arnold-Chiari malformation type I with autonomic dysfunction

Lakesh Kumar Anand[1], Arvind Malhotra[2], Richa Saroa[3],

Sunita Kazal[4], Rashi Sarna[5]

[1]MD, FIMSA, FCCP, MNAMS, FCCS, Professor; [3]MD, Assistant Professor; [4]MD, Senior Resident, Department of Anesthesia and Intensive Care, Government Medical College and Hospital, Sector 32, Chandigarh, (India)
[2]MS, MCh, Associate Professor, Department of Neurosurgery, Government Medical College and Hospital, Sector 32, Chandigarh, (India)
[5]MD, Senior Resident, Department of Anesthesia and Intensive Care, PGIMER, Chandigarh, (India)

Correspondence: Dr. Lakesh Kumar Anand, Professor of Anesthesiology & Intensive Care, Government Medical College and Hospital, Sector 32, Chandigarh 160030, (India); E-mail: lkanand11@gmail.com

ABSTRACT

Arnold–Chiari malformation (ACM) type I with syringomyelia patients can present with autonomic dysfunction. Anesthesia management requires careful preoperative assessment, identification of potential problems and appropriate planning. We present a case report of a

29- years female diagnosed as ACM Type I with syrinngomyelia associated with autonomic dysfunction was operated for foramen magnum decompression. Glidescope was used for intubation to prevent neurological damage associated with neck movement. After placing in prone position patient had hypotension with tachycardia. Supine position was immediately resumed; managed with intravenous (IV) fluid and vassopressure. Patient was again placed in prone position after normalization of blood pressure. Her postoperative period was uneventful.

Key words: Anesthesia; General Anesthesia; Autonomic Nervous System Diseases; Segmental Autonomic Dysfunction; Syringomyelia; Spinal Cord Diseases; Arnold-Chiari Malformation; Arnold-Chiari Malformation, Type I

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INTRODUCTION

Arnold–Chiari malformation (ACM) type I is a disorder of uncertain origin that has been traditionally defined as tonsillar herniation through the foramen magnum. The anomaly is a leading cause of syringomyelia and occurs in association with bony abnormalities at the craniovertebral junction. The natural history is a gradual, stepwise deterioration over many years.[1-4] Type I ACM with syringomyelia may manifest with autonomic dysfunction, often asymptomatic and discovered incidentally on brain or cervical spine MRI scans.[4-7] The anesthetic concerns with general anesthesia are related to the risk of autonomic dysfunction, difficult airway management, damage to the spinal cord and sensitivity to neuromuscular blocking agents. We report the successful management of a patient of syringomyelia with ACM who developed orthostatic hypotension during positioning for foramen magnum decompression.

CASE REPORT

A 29 years female patient weighing 62 kg was scheduled for foramen magnum decompression. She had presented with numbness and weakness of left upper limb for two years and episodes of bilateral frontal headache, giddiness and vertigo for one year. There was no history suggestive of trauma, dysphagia, change in voice and change in bowel or bladder habits.

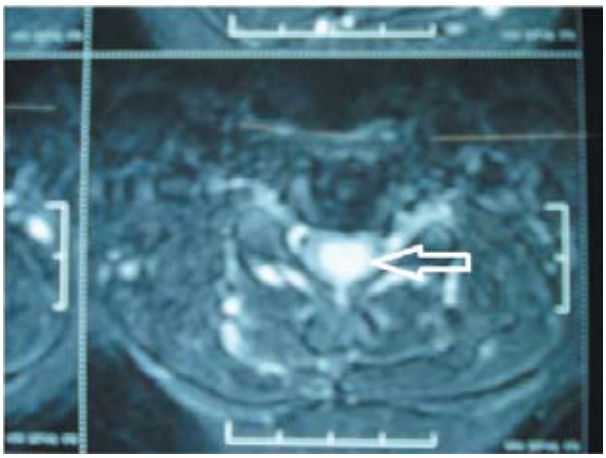

Figure 1: MRI Transverse section of spinal cord, T2 weighted image, arrow showing syrinx in central cord

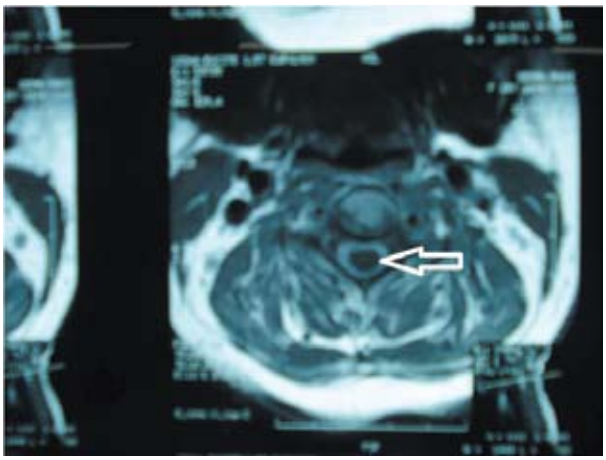

Figure 2: MRI Transverse section of spinal cord, T1 weighted image, arrow showing syrinx in central cord

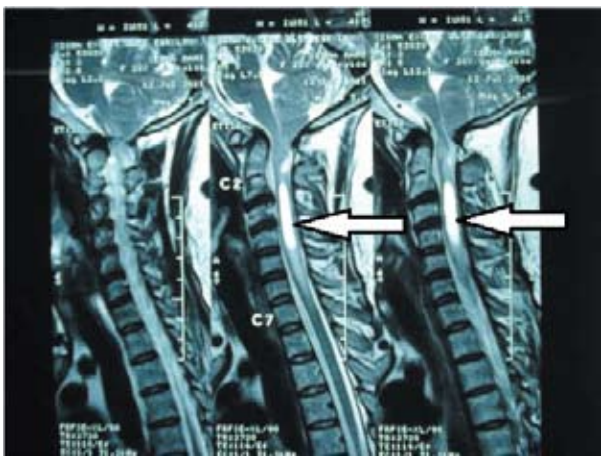

Figure 3: MRI Saggital section of spinal cord, T1 weighted image, arrow showing syrinx in central cord from C2-C6.

On physical examination her pulse rate was 112/min regular, blood pressure sitting 104/70 mmHg and standing 98/68 mmHg right arm. There was reduced sensation to pain and temperature in both hands. She had weakness in both upper limbs (power 4/5) and diminished deep tendon reflexes. Both the lower limbs showed spasticity and weakness (power 4/5). Patient had reduced sweating on palms and dry skin with cuts and lacerations showing glove and stocking type of loss of sensations and autonomic involvement. There was baseline tachycardia and no postural hypotension. Routine preoperative investigations were within normal limits. Preoperative electrocardiography (ECG) showed sinus tachycardia. Chest X-ray was unremarkable.

Her airway assessment showed a Mallampatti grade II and limited neck movements associated with pain and vertigo; a difficult intubation was anticipated. MRI of spine revealed a syrinx in spinal cord extending from C2-C4 with downward herniation of cerebellar tonsils till C2 (Figures 1-3). She was diagnosed as a case of ACM Type I with syringomyelia, and foramen magnum decompression surgery was planned.

Patient was premedicated with oral alprazolam 0.25 mg and ranitidine 150 mg night before and 60-90 min before the expected time of surgery. On the arrival in operating room, an intravenous line was started and inj. glycopyrrolate 0.2 mg was administered IV. Standard monitoring continuous ECG, heart rate (HR), non-invasive blood pressure (NIBP), oxygen saturation (SpO_2) and end-tidal CO_2 ($EtCO_2$) was begun before induction of anesthesia. Anesthesia was induced with inj. fentanyl 100 μg and inj. propofol 100 mg. After checking the ability to achieve adequate mask ventilation, atracurium 0.6 mg/kg was used to facilitate muscle relaxation. Glidescope was used for intubation to prevent neurological damage associated with neck movement, a 7.5 mm ID endotracheal tube was used and placement confirmed by identifying breath sounds plus square-wave capnograph trace. Central venous pressure, invasive blood pressure (IBP) and urine output monitoring were also started. Patient was placed in prone position keeping the head and neck in neutral position to avoid any movement of cervical cord.

After placing in the prone position BP fell to 60/34 mmHg and heart rate rose to 140/min. Supine position was immediately resumed; 500 ml of crystalloid was rushed and inj. phenylephrine 50 μg IV bolus followed by infusion 25-50 μg / min was titrated to mean arterial pressure (MAP) $\approx$ 60 mmHg. After stable hemodynamics patient was again placed in prone position. To our surprise, she again developed hypotension and

tachycardia. In the absence of correctable cause and considering unmasking of autonomic disturbance, phenylephrine infusion was titrated to MAP ≈ 60 mmHg. Anesthesiologist and neurosurgeon decided to continue with the proposed surgical procedure. Anesthesia was maintained using 60% nitrous oxide in oxygen and sevoflurane. Surgery lasted for two and a half hours. Blood loss (≈100 ml) was replaced with colloids. Intraoperatively, patient's vital parameters remained fairly stable (within 20% of the baseline value).

On completion of surgery inj. ondansetron (6 mg) and paracetamol 1 gm IV given to reduce emesis and pain respectively. Residual neuromuscular block was reversed using injection neostigmine and glycopyrrolate combination at the end of surgery. Patient was extubated when awake, breathing spontaneously and responded to verbal command, and shifted to ICU for observation and monitoring. Her postoperative period was uneventful.

DISCUSSION

Type I ACM consists of a downward displacement of the cerebellar tonsils and the medulla through the foramen magnum and spinal canal to compress the spinal cord, consequently causing blockage of the spinal canal and the stoppage of cerebral spinal flow, leading to syringomyelia.[1-3] Syringomyelia is a condition in which a cavity called a syrinx develops in the spinal cord and is filled with cerebrospinal fluid. The pathophysiology of syringomyelia is consistent with the theory that a spinal subarachnoid block increases spinal subarachnoid pulse pressure above the block, producing a pressure differential across the obstructed segment of the spinal subarachnoid space, which results in syrinx formation and progression.[10] Diagnosis of ACM type I is made through a combination of patient history, neurological examination, or discovered incidentally on brain or cervical spine MRI scans. MRI is the diagnostic test of choice for ACM I, since it easily shows the tonsillar herniation as well as syringomyelia, which occurs in 20 to 30 percent of cases.[8-9] Type I ACM patient with autonomic dysfunction can present with postural hypotension, impairment of circulatory reflexes, sinus arrhythmia, postural tachycardia.[7,11-13]

These patients if scheduled for craniotomy already present with some degree of autonomic dysfunction, and significant compression of the neural elements in the craniocervical junction. The anticipated problems to the anesthesiologists during induction, positioning for surgery, maintenance or emergence of anesthesia include autonomic dysfunction, difficulty of airway management and abnormal sensitivity to neuromuscular blocking agents.

Autonomic function should be evaluated in patients with significant brainstem involvement. Subclinical autonomic dysfunction, a well recognized condition in ACM type I, can result in unstable hemodynamic status, lack of compensatory response to hypotension. Invasive monitoring including: CVP for monitoring volume status, IBP for measuring continuous blood pressure should be closely monitored especially if autonomic dysfunction is suspected. The patient should be closely monitored in the immediate 24 hours following surgery.[10] We used CVP monitoring to assess and manage the intravascular volume in case of blood loss. IBP monitoring was started to enables close control of hemodynamic parameters and effectively attenuated cardiovascular response to intubation, a response which could potentially stimulate progression of a syrinx, and hypotension associated with postural change.

The prone position is used for foramen magnum decompression. Head can be positioned in a pin head holder (applied before turning) or a horse shoe head rest. The final position commonly entails neck flexion, reverse Trendelenburg, hands by side of body and elevation of the legs. Pressure should be as evenly distributed as possible over facial structures and especially orbit and nose should be protected. Other pressure points to be checked include the axillae, breasts, iliac crests, femoral canals, genitalia, knees and heels. The anesthesiologist should have a plan for detaching and reattaching monitors in an orderly manner to avoid prolonged monitoring gap, and reverse the position to supine if at all required. We immediately identified the hemodynamic derangement in the form of hypotension and tachycardia, supine position was immediately resumed and patient was managed with bolus of IV fluid and phenylephrine.

In conclusion, ACM Type I and its associated disorders may pose anesthetic risks. Hence, a detailed preoperative examination of autonomic dysfunction especially in cases with cervical syrinx should be done in order to have a better management during surgery. A safe anesthetic management can be achieved by careful attention to the derangements that occur with the disease. Optimal patient outcome will be improved with an interdisciplinary team management including anesthesiology, neurology and neurosurgery services.

Conflict of interest: None declared by the authors

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