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General Anesthetic Management for Foramen Magnum Decompression and Tonsillectomy in a Patient with Arnold-Chiari Malformation Type 1: A Case Report

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Abstract **Case Report**

Arnold-Chiari Malformation Type 1 (ACM-1) is a neuroanatomical anomaly leading to unique anesthesia challenges. We detail the anesthetic management of a 35-year-old female with ACM-1 undergoing foramen magnum decompression and tonsillectomy. Her obesity and tonsillectomy history posed postoperative risks, requiring specialized care. After surgery, she was monitored in the intensive care unit for 24 hours under sedation, then successfully extubated. She was discharged two weeks later without complications. Proper management of ACM-1 patients demands expert collaboration, thorough assessment, and tailored anesthesia planning for surgical safety and success.

Keywords: Arnold-Chiari Malformation Type 1; Foramen magnum; General Anesthesia; Tonsillectomy.

INTRODUCTION

Arnold-Chiari malformation encompasses a group of neuroanatomical abnormalities where a downward displacement of the cerebellar tonsils and occasionally, other structures of the posterior fossa, is evident into the cervical spinal canal [1].

The Arnold-Chiari Malformation Type 1 (ACM-1) is the most common subtype of Chiari malformations, distinguished by a downward herniation of the cerebellar tonsils through the foramen magnum. While it is often diagnosed in adulthood, the anatomical variations and subsequent neurological symptoms - such as headaches, cerebellar dysfunction, and occasionally spinal cord compression - demand tailored approaches in perioperative care, especially when general anesthesia is involved [2].

Administering general anesthesia to patients with ACM-1 is not a straightforward task. Beyond the conventional challenges of anesthesia, these patients present a heightened risk profile due to potential alterations in cerebrospinal fluid dynamics, increased sensitivity to changes in intracranial pressure (ICP), and specific airway management concerns [3]. Ensuring safe anesthesia induction, maintenance, and emergence in ACM-1 patients necessitates a profound understanding of the pathophysiology and its interaction with anesthetic agents and techniques.

We present the anesthetic management for a patient with ACM-1 and tonsillar hypertrophy, scheduled for foramen magnum decompression surgery and tonsillectomy.

CASE PRESENTATION

A 35-year-old female patient (95 kg, 144 cm) came with complaints of numbness in her left upper limb for three months. MRI of spine revealed a syrinx in spinal cord extending from C2-T2 with downward herniation of cerebellar tonsils till C1 (Fig. 1). Main pathological finding was cerebellar tonsillar herniation along with caudal herniation through the foramen magnum (Fig 2). She was diagnosed with ACM-1 and foramen magnum decompression surgery was planned. The patient underwent routine preoperative evaluations, including electrocardiography and laboratory blood tests, which were all within normal limits. Chest X-ray was unremarkable. Tonsillar hypertrophy was incidentally observed during the preoperative

laryngoscopy. A difficult intubation was anticipated. It was decided to also perform a tonsillectomy additionally first.

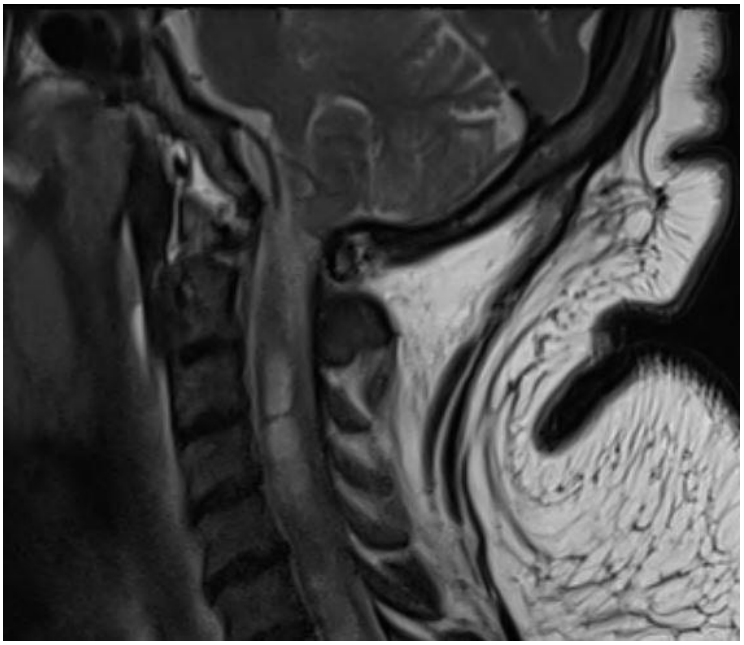

Figure 1: Sagittal T1-weighted MRI shows downward herniation of the cerebellar tonsils through the foramen magnum

Figure 2: Sagittal T2-weighted MRI shows syrinx in spinal cord from C2-T2

Upon entering the operating room, she was monitored with electrocardiography electrodes, non-invasive blood pressure, pulse oximetry, bispectral index. Initial vital signs were all within normal range. Anesthesiologists and neurosurgeons discussed the possibility of increasing ICP due to anesthetic and surgical procedures. Since the corrective surgery for this patient was planned to relieve the increased ICP associated with ACM-1, we decided not to conduct ICP monitoring for this patient. Instead, we opted to focus

on strict blood pressure and CO2 partial pressure monitoring to watch for increases ICP.

Anesthesia was induced with a bolus of lidocaine-50mg and propofol-150mg, and remifentanil-infusion at 0.11mcg/kg/min. After checking the ability to achieve adequate mask ventilation, rocuronium 0.6 mg/kg was used to facilitate muscle relaxation. Rigid video laryngoscope (Reusable Rigid Video Stylet, Novamed) was used for intubation to be expected difficult airway due to tonsillar hypertrophy and to prevent neurological damage associated with neck movement. After confirming deep neuromuscular block through train-of-four, an attempt was made for endotracheal intubation. Due to significant bilateral tonsillar hypertrophy, the vocal cords were not readily visible initially. With utmost caution, a Rigid Intubation Scope was introduced to identify the vocal cords (Fig. 3), and intubation was carried out successfully without complications. 7.0-mm inner-diameter-cuffed reinforced endotracheal tube (Shiely^{TM} Lo-Contour, Covidien Ireland Limited) was used, and placement confirmed by identifying breath sounds. Central venous pressure, invasive radial arterial blood pressure (IBP) and urine output monitoring were started. The patient was kept on ventilation mode with volume control mode, and tidal volume was set at 8 mL/kg, respiratory rate at 14 breaths per minute, and FiO2 at 50%. Anesthesia was maintained with sevoflurane 2.1 vol% in 50% oxygen-enriched air and remifentanil 0.2 μg/kg/min.

Figure 3: The Rigid video laryngoscope finding during the intubation shows a narrow airway due to tonsillar hypertrophy

For the initial surgery, a tonsillectomy was performed in the supine position by the otolaryngologist, and the procedure was carried out with the neck in a state devoid of any cervical hyperextension. The procedure lasted for a duration of one hour, and it concluded smoothly without major bleeding. In preparation for the second neurosurgical procedure, foramen magnum decompression surgery, the patient's position was changed to the prone position. Care was taken to monitor for any increase in IBP and to avoid excessive neck extension during the repositioning. Given that the surgery involved the use of head pins, remifentanil was administered to ensure adequate pain control. The foramen magnum decompression surgery proceeded for an approximate duration of 8 hours. A hemorrhage of around 450cc occurred and was subsequently managed by the administration of 2000cc of crystalloid fluid. Vital signs remained stable throughout the procedure. During the surgery, we performed an Arterial Blood Gas Analysis every hour to ensure that the paCO2 levels were maintained at an appropriate level. The surgery proceeded as planned and was completed without any complications.

The anesthesiologist and neurosurgeon decided to maintain endotracheal intubation during postoperative care. This decision was based on the assessment that extubation posed a substantial risk. The primary justification pertains to the patient's pronounced obesity, a substantial high-risk factor. The secondary justification emanates from the envisaged complexities in airway management due to the patient's antecedent history involving tonsillectomy and cervical spine surgery. Immediately following the surgery, the patient was transported to the Intensive care unit while being maintained in an intubated and sedated state. Subsequently, a 24-hour period encompassing ventilator care and pain control was conducted. After 24 hours, the patient regained consciousness, exhibited

appropriate responsiveness, and did not complain of severe pain. Consequently, extubation was carried out, and the patient was transferred to the general ward. Two weeks later, she was discharged without significant complications.

DISCUSSION

ACM-1 is a complex congenital condition characterized by structural anomalies involving the cerebellum and brainstem. This malformation commonly presents with hydrocephalus, and other neurological abnormalities [4]. Given the intricate nature of this condition, the administration of general anesthesia requires careful consideration and planning. Patients with ACM-1 are susceptible to brainstem and cerebellar herniation, which may compress essential neural structures. Their surgical treatment often aims to relieve such compression and handle other issues like hydrocephalus [5].

The presented case highlights a complex scenario involving a female patient diagnosed with ACM-1 who underwent subsequent surgical interventions. This case offers valuable insights into harmonizing patient-specific physiological considerations with the expertise of the medical team.

Airway management in ACM-1 patients requires meticulous planning due to potential anatomical complications, like a restriction of cervical movement. An anesthesiologist should be equipped and ready for airway challenges [5]. In this case, due to the presence of ACM-1 combined with tonsillar hypertrophy, a difficult airway was anticipated. Therefore, we prepared a video scope in advance and, by preparing for potential challenging airway management situations, we successfully performed the intubation without any issues.

For patients with ACM-1, monitoring ICP during anesthesia is essential, though it often demands invasive approaches. The primary goal is to maintain cerebral blood flow while stabilizing ICP [6]. Despite the significance of direct ICP monitoring, it entails highly invasive techniques. In this case, instead of using direct ICP monitoring, rigorous control of IBP and PaCO2 was emphasized during the surgery to mitigate the risk of increased ICP.

The decision to employ the prone position during surgery was pivotal, given the intricate nature of the procedure and the imperative of maintaining the patient's safety and accuracy. Changing positions during surgery can cause stress to a patient, which can lead to an increase ICP. Therefore, appropriate pain control and a gentle change of position were necessary. Effective positioning management held the potential to influence surgical success and patient safety significantly [7].

During the recovery from anesthesia, measures should be in place to prevent events that could raise ICP, ensuring a controlled and closely monitored extubation process. In this case, due to severe obesity and the combination of tonsillectomy and cervical spine surgery, the patient did not awaken from anesthesia and was transferred to the Intensive care unit under sedation for further care. It's crucial to formulate a comprehensive treatment plan that ensures safety during general anesthesia by considering each patient's physiological characteristics and medical history.

This case highlights the significance of patient-centered care and the collaboration and expertise of the medical team. The intricacies of surgery and general anesthesia necessitate a prioritization of patient safety through seamless teamwork and the establishment of a harmonious treatment plan. The medical team must conduct a thorough assessment and formulate an effective treatment plan, keeping in mind the patient's unique physiological attributes and medical history.

CONCLUSION

A patient with ACM-1 presents unique challenges to anesthesiologists due to their complex neurological anatomy. A comprehensive understanding of the underlying pathology, careful preoperative assessment, meticulous airway management, ICP monitoring, and tailored anesthetic plans are essential for ensuring the safety and successful outcomes of surgical procedures in this patient population. Collaboration among neurosurgeons, anesthesiologists, and the medical team is crucial for the optimal care of these patients.

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