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Successful Cesarean Section Under General Anesthesia in a Patient with Chiari 1 Malformation and Temporo-Mandibular Joint Dysfunction: A Case Report

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Abstract: *Background*: Chiari malformation is a group of congenital abnormalities involving the cerebellar tonsillar herniation through the foramen magnum which affects the relationships between the cerebellum, brainstem, upper cervical cord, and the base of the cranium. CM-I (Chiari Malformation 1), is seen in adult patients and is associated with cranio-cervical abnormalities. Most of the patients with CM-I get symptoms when a cerebellar tonsillar herniation is greater than 5 mm. *Case Information*: This case report highlights the anaesthetic management of Chiari malformation with 8 mm cerebellar tonsillar descent in a pregnant patient with known difficult airway due to concurrent Temporo-mandibular joint dysfunction, who presented to the hospital for an elective cesarean section. *Discussion*: Childbearing women with Type I CM have concerns related to an increased cranial CSF pressure during pregnancy and labor. This difference in pressure above and below the foramen magnum may become worse following a lumbar puncture or spinal injection for spinal anesthesia and may lead to worsening of the cerebellar herniation leading to strangulation of the brainstem. These risk factors affect the choice of anaesthetic in these patients. *Conclusion*: As shown in this case report, the multidisciplinary discussion raised concerns with competing safety concerns of difficult airway if given general anesthesia and the risk of brainstem herniation with neuraxial anesthesia, despite evidence showing infrequent complications regardless of choice of anesthetic. Careful administration of the anesthetic technique, general or regional anesthesia in this group of patients can lead to favorable outcomes following multi-disciplinary discussion.

Keywords: Chiari 1 Malformation, Temporo-Mandibular Dysfunction, Pregnancy, General Anesthesia, Difficult Intubation

1. Introduction

Chiari malformations are a type of anomaly where the lower end of the cerebellar tonsils herniates beyond the foramen magnum [1]. There are 4 types of Chiari malformations. This Classification is based on the extent of the severity of malformations [2]. The most common with a prevalence of 0.7% is the Chiari I malformation which has an adult-onset and consists of a 5 mm descent of the caudal tip bulging through the foramen magnum [3]. It is often asymptomatic but may present with headaches, neck pain, or ataxia. The anesthetic concerns in women with Type-I ACM (Arnold Chiari type 1 Malformation) are related to an increase in cerebrospinal fluid (CSF) pressure associated with pregnancy and the second stage of labor [4], or a differential effect between cranial and spinal CSF pressure. The anesthetic and obstetric management of parturient presenting for delivery remains controversial. According to Gruffi et al anesthetic complications are uncommon in patients with ACM-I regardless of the type of anesthesia administered for these patients [2]. Although anesthetic and obstetric care is planned according to institutional preferences, personal experience of the anesthetic procedure has led to the successful management of these patients. Recent studies also reiterate the importance of a multidisciplinary approach in the management of these patients which has been shown to improve outcomes. We describe successful anesthetic management of a pregnant

patient with worsening of symptoms during pregnancy and concurrent difficult intubation secondary to poor mouth opening due to 3 previous mandibular surgeries for odontogenic keratocyst of right mandible. Using the Obstetric Anaesthetists' Association and Difficult Airway Society guidelines for optimizing safe general anaesthetic technique, we planned a framework for airway management in this patient in our clinical setting [5].

2. Case Report

A 38-year-old female G4 para 3+0 presented at 34 weeks and 1-day gestation with long-standing migraines with aura, partial bilateral hearing loss, and astigmatism since childhood. The migraines were progressively worsening during this pregnancy and were exacerbated by Valsalva maneuvers. She had no focal neurological deficits and had no prior history of convulsions.

She reported occasional breathlessness at night. No cough, no chest pain, no peripheral edema, and normal exercise tolerance. She had no history of nausea, vomiting and reported normal bowel movements and urinary functions. A few years prior, she was diagnosed with odontogenic keratocyst of the right mandible and had undergone three serial mandibular reconstruction surgeries. Following the above surgeries, she developed Left Trigeminal Neuralgia and reduced temporomandibular joint function which resulted in limited mouth opening (could not open the mouth beyond 2 fingers breadth). Prior to this scheduled surgery, she underwent a laparoscopic appendectomy under general anesthesia with a Maxillo-facial surgeon in attendance during intubation and extubation due to TMJ function concerns. She had no anesthetic complications in any of the above surgeries. She was a non-smoker, denied use of alcohol, and was allergic to peanuts. Her antenatal profile was unremarkable. Dating scan at 12 weeks 3 days showed normal early anatomy and EDD which correlated well with the dates. Fetal ultrasound anomaly scan showed normal anatomy, and the placenta was posterior. She had a normal Oral Glucose Tolerance Test and blood pressure during this pregnancy. All her previous three labors were spontaneous in onset at term with precipitated labor and vaginal deliveries. There were no major delivery complications although she did report worsening headaches with each delivery. A prior MRI done a year ago showed a Chiari I malformation with 6 mm inferior descent of the cerebellar tonsils below the foramen magnum. A repeat MRI done during pregnancy showed an increased descent of the cerebellar tonsils at 8mm.

A multidisciplinary discussion involving the consultant obstetrician, neurosurgeon, neonatologist, midwife, and anesthesiologist was held. Following risks related to pregnancy, delivery and anesthesia were identified:

1) Risk of Chiari progression due to increased ICP from uterine contractions, labor pain, and Valsalva maneuver which may result in respiratory arrest.
2) Risk of worsening cerebellar herniation in case of inadvertent CSF leak after epidural or spinal analgesia/ anesthesia attempt.
3) Anticipated difficult intubation due to limited mouth opening secondary to mandibular surgeries in case of an emergency cesarean section and pregnancy.
4) Risk of aspiration due to pregnancy.
5) Risk of brainstem compression in case of neck hyperextension during intubation
6) Risk of early and precipitate labor from the previous history.

Risks versus benefits of a general versus regional anesthetic were discussed at length with the patient and her husband. The agreed delivery plan, after giving her steroids for fetal lung maturation, was an elective cesarean section at 37 weeks gestation to prevent worsening of the intracranial pressure due to labor pain. The anesthetic plan included general anesthesia following awake fiberoptic intubation technique in view of the anticipated difficult airway.

The patient was reviewed in the anesthesia preoperative clinic 2 weeks prior to the surgery with normal vitals and BMI of 27.2 kg/m^2. During airway examination, we found a short mobile neck, Mallampati classification of III, and inter-incisor distance of 2 finger breadth. We discussed anesthetic concerns related to securing her airway with focus on fiber-optic intubation as the safest mode of choice and she consented to general anesthesia with this plan.

Pre-operative laboratory tests included a full hemogram and urea, electrolytes, and creatinine which were all within normal ranges. The parturient was admitted the night prior to her planned Cesarean section. Routine CTG done in the morning demonstrated fetal tachycardia which resulted in a category 2 emergency Cesarean section.

In the operating room, compression stockings for thrombo-prophylaxis were attached, and the patient was connected to the standard ASA monitoring including pulse oximetry, non-invasive blood pressure, and an electrocardiogram. Intravenous access was established with two 18 gauge cannulas on her left hand.

Due to the emergent nature of surgery, fiberoptic intubation apparatus was unavailable at that time; thus, video laryngoscope and McCoy blades were prepared for difficult intubation. The patient was pre-oxygenated for more than 3 minutes with face mask with 100 percent oxygen. Intravenous metoclopramide 0.1mg/kg, remifentanil 1mcg/kg, propofol 2mg/kg and atracurium 0.5mg/kg were administered for induction of anaesthesia. The first intubation failed with a video laryngoscope with a successful second attempt using McCoy laryngoscope without extending her neck and maintaining the cervical spine in line stabilization. After the correct position of the endotracheal tube was confirmed by directly visualizing the endotracheal tube passing through vocal cords, presence of end-tidal carbon dioxide waveform, chest rise, and bilateral breath sounds, the obstetrician was prompted to begin the surgery.

Intraoperatively, she was positioned supine and bed was tilted to the left for the prevention of supine hypotension and active warming using a bair hugger was initiated. She was maintained on isoflurane/oxygen/air mixture to a MAC of 1.

Normocapnia and normoxia were maintained throughout the surgery with a peak airway pressure of 15-18 mmHg. She was hemodynamically stable and an estimated blood loss of 800 ml was recorded at the end of surgery.

Analgesia included parenteral Paracetamol, Diclofenac and ultrasound guided bilateral Transversus abdominis plane (TAP) block. The patient was extubated awake; however, she developed a severe laryngospasm which was managed using airway maneuvers and head up position without of extension of her cervical spine. Post-extubation neurological evaluation was at baseline; no neurological deficits were noted.

Post-operatively, she was admitted to High Dependency Unit and orders involved adequate analgesia and hydration, nurse head up, close neurological monitoring. The post-operative recovery was unremarkable. She reported a mild headache VAS 2/10 which was relieved by Paracetamol. She had no new focal neurological deficits, remained alert and oriented.

She was discharged on post-operative day 4 and referred to neurosurgery clinic for follow -up where a conservative approach was advised to the CM-I.

3. Discussion

CM-I (Chiari Malformation 1), a congenital anomaly, is seen in adult patient and associated with cranio-cervical abnormalities. Most of the patients with CM-I get symptoms when cerebellar tonsillar herniation is greater than 5 mm and mostly symptomatic when it is more than 12 mm [6].

Childbearing women with Type I CM have concerns related to an increased cranial CSF pressure due to cerebellar herniation during pregnancy and labor, which can lead to the difference in pressure between cranial and spinal CSF. This difference in pressure above and below the foramen magnum may become worse following a lumbar puncture or spinal injection for spinal anesthesia and may lead to worsening of the cerebellar herniation leading to strangulation of the brainstem.

The choice of general anesthesia versus regional anesthesia poses individual risks. Regional anesthesia may lead to tonsillar herniation especially in patients with signs of increased intracranial pressure. General anesthesia in our case posed significant risk in intubation due to difficult airway secondary to the previous mandibular surgeries, extension of the neck during intubation, positioning and reversal.

There are no consensus guidelines on the anesthetic management of patients with CM-I. Acute deterioration of occult CM-I has been described in parturients [7]. On the contrary, the safe use of general anaesthesia (GA) and spinal anesthesia has been documented with known CM-I in parturients [8, 9]. Uneventful spinal anaesthesia with narcotics added to local anaesthetic has been described in a known CM-I parturient for cesarean delivery [10]. However, the degree of tonsillar herniation was not described in the literature. There is limited data on the use of neuraxial techniques in patients with surgically corrected Chiari malformation. Though successful spinal anaesthesia for caesarean delivery in a woman with a surgically corrected CM-I has been described but the authors did advise a word of caution [9].

Gruffi et al concluded that anesthetic complications occur infrequently in patients with Chiari malformation regardless of anesthetic management [2].

The choice of regional or general anesthesia should be individualized using a multidisciplinary approach in the management of the patient. We opted to have general anesthesia due to the increased risk of cerebellar tonsillar herniation following a spinal block because of her worsening symptoms during pregnancy. Although these are rare disorders with significant potential morbidity, labor can be managed by either mode of delivery with careful patient selection.

4. Conclusion

In patients with multiple anesthesia-related risk factors and pre-existing medical conditions, a multi-disciplinary approach improves patient outcome, treatment, and satisfaction. As shown in this case report, the multidisciplinary discussion raised concerns with competing safety concerns of difficult airway if given general anesthesia and the risk of brainstem herniation with neuraxial anesthesia, despite evidence showing infrequent complications regardless of choice of anesthetic.

We achieved successful intubation with endotracheal tube using McCoy blade without extending neck, in management of this parturient with CM-1 (cerebellar tonsillar herniation greater than 8 mm) with anticipated difficult intubation due to limited mouth opening, and increased risk of aspiration during pregnancy. Of note, difficult intubation patients undergoing surgery require experienced physicians who are competent in fiberoptic intubation and difficult airway algorithm. Appropriate doses of remifentanil, propofol, and atracurium led to the successful anesthetic management of this patient for emergency cesarean section. Patients without any neurological symptoms should not undergo sub-occipital decompression for Chiari I malformation during pregnancy. While patients with significant neurologic symptoms prior to conception should be considered for decompression prior to pregnancy [11]. Offering normal vaginal delivery with effective analgesia, for women with Chiari malformation, appears to be safe. A multi-disciplinary team including Obstetrician, Anesthetist, and Neurosurgeon with experience in managing Chiari malformation can provide good pregnancy care [12]. It is very important to carefully select the anesthetic technique for the delivery of a woman with an Arnold-Chiari malformation [13]. According to TEO MM, multidisciplinary discussion and early anesthetic consult are extremely important in determining the outcome of the patient, and that carefully administered spinal anesthesia can be a safe anesthetic option [14]. Waters GFR et al suggested that the mode of delivery for patients with Chiari I malformation and without any signs of raised intracranial

pressure should be based on obstetric considerations. The successful administration of the epidural and spinal anesthesia without any complications in patients with Chiari-1 malformation suggests that these procedures can be carefully performed for these patients [15].

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