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Case Report

Pregnancy with Chiari malformation type II: A Case Report and Literature Review

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Abstract:

This article reports the diagnosis and treatment process of a pregnant patient with Chiari malformation type II. The patient was at 38^{+4} weeks of gestation, complicated by chronic hypertension with preeclampsia and diabetes. A multidisciplinary approach was adopted, and cesarean section under general anesthesia was chosen to terminate the pregnancy, resulting in favorable maternal and neonatal outcomes. Combined with a literature review, the article emphasizes the importance of multidisciplinary management and individualized anesthesia strategies for pregnant patients with Chiari malformation type II.

Key words: chiari malformation type II; pregnancy; cesarean section; multidisciplinary management

Introduction

Chief Complaint and Present Illness: A 31-year-old female, self-employed, at 38^{+4} weeks of gestation, was admitted on June 25, 2024, at 11:39 due to "high blood pressure for 7 months, worsening in 1 day." High blood pressure (139/91 mmHg) and proteinuria (+) were detected during routine prenatal checkup at 15 weeks of gestation. She was prescribed labetalol hydrochloride 100 mg twice daily. Fasting blood glucose was 7.93 mmol/L, but blood glucose was neither controlled nor monitored during pregnancy. Recent glycated hemoglobin (HbA1c) was 6.6%. At 20 weeks of gestation, nifedipine sustained-release tablets 30 mg twice daily were added due to poorly controlled blood pressure, which was then maintained below 140/90 mmHg. She received prenatal care at county and municipal hospitals. One day before admission, her blood pressure rose significantly to 180/110 mmHg, with proteinuria (+++), but no headache, dizziness, or blurred vision. She was hospitalized at a municipal hospital for pregnancy termination. Due to her short neck, a cranial magnetic resonance imaging (MRI) was performed, revealing cervical syringomyelia, cervical meningocele, and cerebellar tonsillar herniation, suggesting Chiari malformation type II. Due to anticipated anesthesia difficulties, she was transferred to our hospital.

Physical Examination on Admission: Menstrual cycle of 30-41 days, with a period lasting 5-7 days. Gravida 2, Abortion 1, Para 0. The last menstrual period was on September 15, 2023. Based on the ultrasound at 12 weeks of pregnancy, the last menstrual period was estimated to be September 29, 2023, with an expected delivery date of July 5, 2024.

Auxiliary Examinations: Cranial MRI (June 24, 2024): Cervical syringomyelia, cervical meningocele, and cerebellar tonsillar herniation, consistent with Chiari malformation type II.

Admission Diagnosis:

Intrauterine pregnancy at 38^{+4} weeks, first fetus, LOA, no labor signs.

Pregnancy with Chiari malformation type II.

Chronic hypertension complicated by preeclampsia.

Diabetes complicating pregnancy.

Multidisciplinary Consultation and Decision-Making: After admission, the patient was given Labetalol, 100 mg Q8H and Nifedipine sustained-release tablets, 30 mg BID, along with intravenous infusion of Urapidil for blood pressure control and intravenous Magnesium Sulfate for antispasmodic treatment. Considering the patient's critical condition, an immediate multidisciplinary consultation was organized by the Medical Affairs Department, involving neurosurgery, anesthesiology, neurology, neonatology, otolaryngology, intensive care medicine, and cardiology. The neurosurgeons and neurologists believed that the patient currently had no muscle strength impairment, normal daily activities, and no incontinence. However, during delivery, there was a risk of significant changes in circulation and intracranial pressure, as well as large blood pressure fluctuations, which could exacerbate her condition. Spinal anesthesia might affect intracranial pressure changes, and the risks and benefits were weighed to select the anesthesia method. The anesthesiologists, in consultation with the neurologists, considered that spinal anesthesia carried a high risk due to the spinal cord abnormalities and proposed general anesthesia. Given the patient's short neck, there was a possibility of difficult intubation, failure of intubation, and hypoxemia, which could affect the mother and fetus. The otolaryngologists prepared for possible tracheotomy due to the patient's obesity. The cardiologists suggested that the patient's complex condition and high blood pressure required an investigation into the cause of hypertension, possibly including tests for hypertension triad, supine and standing positions, renal artery Doppler, and cardiac MRI to assess cardiac lesions and vascular

abnormalities. They recommended adjusting antihypertensive medications as necessary, continuing intravenous Urapidil for blood pressure control, and proceeding with urgent cesarean section. Based on the opinions of all departments, it was decided to perform an emergency cesarean section, and preparations were made accordingly.

Surgical Process and Postoperative Management: On June 25, 2024, at 16:55, the patient underwent a lower segment cesarean section and bilateral uterine artery ligation under general anesthesia (combined intravenous and inhalation anesthesia). The patient was placed in the supine position, and the abdomen was disinfected and draped. A transverse incision was made three transverse finger widths above the pubic symphysis. The abdominal fat was thick, and the lower segment of the uterus was well-formed. The peritoneum was opened, exposing the lower segment of the uterus. A 2 cm incision was made in the myometrium of the lower segment of the uterus, and the uterine muscle layer was bluntly dissected upward in an arc shape. After amniotomy, the amniotic fluid was clear, with a volume of approximately 900 ml. The fluid was suctioned out, and at 17:05, a male neonate was delivered in the LOA position with assistance. The neonate's airway was cleared; the umbilical cord was clamped and handed over for further processing. The neonate was warmed on a radiant warmer, intubated, and given positive pressure ventilation with a resuscitation bag. The umbilical cord was wrapped with sterile gauze. The neonate weighed 3,970 g, with no obvious malformations. Apgar scores: 5 (1 min), 8 (5 min)5, 8 (10 min). The neonate was transferred to the neonatology department for further treatment. After the delivery of the fetus, Oxytocin 10 U was administered intravenously. The uterus contracted poorly, and there was slightly more bleeding from the uterine cavity. Uterine massage and bilateral artery ligation were performed due to poor contraction. The bladder was further pushed down, and the bladder side space was freed. The bilateral uterine arteries were ligated with No. 1 absorbable sutures 2 cm below the level of the incision. The bleeding improved significantly, and the uterine contraction improved. The placenta and membranes were delivered intact. The uterine cavity was cleaned with a wet gauze, and no abnormalities were found in the appearance of the uterus and its bilateral adnexa. The uterine incision was not extended. The uterine myometrium was sutured continuously with No. 1 absorbable sutures, and the outer 1/3 of the myometrium was buried in parallel with a mattress suture. The peritoneum was closed intermittently, and the surgical field was irrigated with normal saline. No bleeding was found upon examination of the surgical site. The surgery went smoothly, with an estimated blood loss of 460 ml during the operation. Considering the patient's critical condition, she was transferred to the intensive care unit. Postoperatively, she was given antispasmodics, antihypertensives, antibiotics respiratory support, oxytocin to promote uterine contraction, and fluid replacement. The patient was transferred back to our department on the second day after surgery when her vital signs were stable. Her condition improved, and she was discharged at her request on the fifth day after surgery. She is currently under close follow-up.

2. Discussion

Chiari malformation type II: Arnold–Chiari malformation, also known as Chiari malformation, is a common congenital developmental abnormality. In 1891, Austrian pathologist Hans Chiari first reported in detail a case of cerebellar tonsillar ectopia in the craniovertebral junction area. It is caused by abnormal embryonic development that causes the lower part of the cerebellar tonsils to descend below the foramen magnum into the cervical canal. In severe cases, parts of the lower medulla oblongata and the lower part of the fourth ventricle also herniate into the craniovertebral junction area, forming a group of heterogeneous anatomical abnormalities [1]. It is often associated with syringomyelia and can cause obstructive hydrocephalus due to impaired cerebrospinal fluid (CSF) circulation. It is also frequently accompanied by other craniovertebral abnormalities such as meningocele, spina bifida, and cerebellar hypoplasia. Pathologically, there are four types, with Type II being the most common [2]. Chiari malformation type II is characterized by downward displacement of the cerebellar vermis, partial brainstem, and fourth ventricle into the foramen magnum and upper cervical canal, often accompanied by syringomyelia and hydrocephalus. The clinical symptoms are caused by compression of the spinal cord and brainstem by the herniated brain tissue, including headache, weakness in the upper limbs, decreased pain and temperature sensation in the shoulder and arm, dysphagia, dizziness, nausea, ataxia, and even paralysis. The severity of clinical symptoms varies. The exact cause is not clear, but it is believed to be related to multiple factors, mainly gene deletion [3]. The gold standard for diagnosis is currently magnetic resonance imaging (MRI); the presence of cerebellar tonsils more than 5 mm below the foramen magnum on MRI can make the diagnosis [1]. MRI findings in Chiari malformation type II include a small posterior fossa, downward displacement of the cerebellar tonsils through the foramen magnum, associated infratentorial and supratentorial abnormalities, downward displacement and elongation of the medulla and fourth ventricle, and intracranial deformities and hydrocephalus [4]. The treatment of Chiari malformation requires multidisciplinary collaboration, with the goal of alleviating symptoms and preventing further deterioration. In 1950, Gardner and Goodall first published a series of studies on surgical treatment of Chiari malformation and syringomyelia [5]. The most widely used surgical treatment is craniovertebral junction decompression [6], which restores normal CSF flow and relieves pressure on the brainstem and cerebellum [7]. Syringomyelia and Chiari syndrome are very rare diseases. With the increased use of MRI, more cases have been identified. Some cases are asymptomatic and require standardized definitions, diagnosis, and treatment [8]. The patient in this case had no clinical symptoms, had regular prenatal checkups, and was diagnosed with Chiari malformation type II in the late pregnancy due to abnormal cranial and cervical morphology on MRI.

Challenges of Pregnancy with Chiari malformation type II: Chiari malformation with syringomyelia most commonly occurs in the cervical and thoracic regions, and most patients exhibit signs and symptoms related to syringomyelia before delivery [9]. However, there are reports suggesting that syringomyelia may spontaneously regress during pregnancy and childbirth. This is because syringomyelia may be caused by physiological CSF flow disturbances in the craniovertebral junction and around the skull. During pregnancy, the increased intra-abdominal pressure due to the growing fetus, Valsalva maneuvers during childbirth, and hemodynamic changes to accommodate pregnancy can alter CSF flow, leading to the disappearance of syringomyelia. Therefore, it is important to continue regular monitoring of syringomyelia during pregnancy and the postpartum period [10, 11]. It is also suggested that if symptoms related to Chiari malformation occur during pregnancy, surgical treatment can be considered in the mid-pregnancy period [12].

Importance of Multidisciplinary Collaboration: Considering the risk of poor neurological prognosis during pregnancy and the risk of increased intracranial pressure during childbirth, the management of Chiari malformation during pregnancy is challenging and should be provided by a multidisciplinary team with experience in treating Chiari malformation [13].

Mode of Delivery: The mode of delivery is not necessarily changed due to Chiari malformation. Most women choose their preferred mode of delivery, either normal vaginal delivery or cesarean section. It is also believed that effective analgesia for normal vaginal delivery is safe for patients with Chiari malformation [14]. Although there are concerns that vaginal delivery may worsen syringomyelia, there have been no reports of any long-term neurological deterioration due to the mode of delivery [9]. Some scholars have suggested that Chiari malformation significantly increases the cesarean section rate [15]. In this case, the patient was at term and had chronic hypertension complicated by preeclampsia with poor blood pressure control. Considering the patient's overall condition, cesarean section was chosen as the mode of delivery.

Controversy and Individualized Strategy for Anesthesia: The choice of anesthesia for pregnant women with Chiari malformation is crucial [16]. Multidisciplinary management and early anesthesia consultation play an important role in the outcome of pregnancy [17]. Some scholars believe that the anesthesia for cesarean section in pregnant women with Chiari malformation can be general anesthesia, spinal anesthesia, or epidural anesthesia. No neurological sequelae have been reported. No adverse neurological consequences were observed at discharge, and it is also believed that effective analgesia for vaginal delivery is safe [15]. However, some studies have suggested that neuraxial analgesia may increase the risk of bleeding and seizures, so epidural anesthesia should be used cautiously [18], and general anesthesia for cesarean section is recommended [19]. There are also reports of pregnant women with Chiari malformation and severe pregnancy-induced hypertension who had cesarean sections under epidural anesthesia due to difficulties with tracheal intubation under general anesthesia [20]. Therefore, the choice of anesthesia should be individualized based on the experience of the anesthesiologist and the patient's condition. In this case, after multidisciplinary consultation, it was recommended to perform cesarean section under general anesthesia. The possibility of difficult intubation was considered before surgery, and the otolaryngology department was asked to prepare for possible tracheotomy. However, the intubation went smoothly during anesthesia, the surgery went well, and the patient recovered smoothly after surgery.

3. Conclusion

The management of pregnancy complicated by Chiari malformation type II requires multidisciplinary collaboration, with a focus on blood pressure control, anesthesia risks, and timing of delivery. Individualized selection of anesthesia and mode of delivery can reduce the risk of maternal and fetal complications. Regular follow-up of syringomyelia changes is crucial for prognosis assessment.

Ethical Approval and Consent: This study was approved by the hospital's ethics committee, and the patient provided informed consent.

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