BLOQUEIO SUBARACNOIDEU NA MALFORMAÇÃO DE CHIARI TIPO 1 – DESCRIÇÃO DE CASO CLÍNICO

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Palavras-chave:
- Anestesia Obstétrica;
- Bloqueio Subaracnoideu;
- Cesariana;
- Malformação de Arnold-Chiari

Resumo

A malformação de Chiari tipo 1 caracteriza-se por uma herniação das amígdalas cerebelosas abaixo do nível do buraco magno. O quadro clínico é variável, não existindo consenso relativamente ao manuseio obstétrico e anestésico destes doentes. Apresentamos o caso de uma grávida com malformação de Chiari tipo 1 não corrigida, submetida a cesariana sob bloqueio subaracnoideu. A paciente referiu posteriormente rigidez cervical ligeira, autolimitada, sem cefaleia. A literatura é escassa, com referência a apenas dois casos de grávidas submetidas a cesariana sob bloqueio subaracnoideu. Com a apresentação do caso pretendemos discutir a segurança deste tipo de anestesia no contexto clínico mencionado.

IS SPINAL BLOCK SAFE FOR CESAREAN DELIVERY? – A CASE REPORT OF A PATIENT WITH UNCORRECTED TYPE 1 CHIARI MALFORMATION

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- *Anesthesia, Spinal;*
- *Arnold-Chiari Malformation;*
- *Cesarean Section*

Abstract

Type 1 Chiari malformation is a downward displacement of the cerebellar tonsils with hindbrain overcrowding. The clinical presentation is highly variable and obstetric and anesthetic management is not consensual. We report the case of a patient with uncorrected type 1 Chiari malformation presenting for elective cesarean delivery in whom a spinal block was performed. Anesthesia and surgery were uneventful. Mother's sole complaint was neck stiffness which subsided after 3 days. Literature is scarce, reporting only 2 previous uncorrected type 1 Chiari malformation patients submitted to spinal anesthesia for cesarean delivery. The safety of spinal anesthesia in this scenario is discussed.

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BACKGROUND

Type 1 Chiari malformation (CM-1) is a neurologic condition resulting from an underdeveloped posterior cranial fossa producing a downward displacement of the cerebellar tonsils below the level of the foramen magnum, with overcrowding of the hindbrain and obstruction to cerebrospinal fluid circulation.[1] In 40-70% of patients there is also associated syringomyelia. The clinical presentation varies from asymptomatic to symptoms resulting from raised intracranial pressure, cranial nerve or bulbar compression or cerebrospinal fluid disturbances, strain-related spinal headache being the most common. Other features include neck stiffness, dizziness, syncope, limb paresthesia or episodic nausea and vomiting.[1] The severity of symptoms does not seem to be directly related to the degree of tonsillar herniation but with the degree of obstruction to cerebrospinal fluid circulation.[2] CM-1 is frequently undiagnosed, either because the patient is asymptomatic, or due to unspecific symptoms. The diagnostic criteria imply brain magnetic resonance imaging (MRI) and the estimated prevalence is 0.1-0.5%,[2] being higher amongst women. The only definitive treatment is decompressive surgery, most commonly through the posterior fossa with dural opening.[2] Concerning the obstetric and anesthetic management of these patients, it is not consensual whether to perform a vaginal or cesarean delivery or if a general or neuraxial anesthesia is the best choice. Any of these options may, through different mechanisms, increase the pressure gradient between the cranial and spinal compartments, thus triggering brain herniation and worsening neurologic symptoms.

Through the report of this case, we discuss the safety of spinal anesthesia in selected uncorrected CM-1 patients presenting for cesarean delivery, a topic scarcely referred in the literature.

The patient has reviewed the case report and gave written consent for publication.

CASE DESCRIPTION

We report the case of a 34-years-old, ASA III primigravida, 97 kg and 169 cm tall, presenting at 39 weeks of gestational age for elective cesarean delivery. At the age of 18 years, the patient was diagnosed with CM-1, presenting as episodes of syncope during heavy exercise when combined with stressful periods of sleep deprivation. At the time neurosurgical correction was proposed but the patient refused it; at present she is minimally symptomatic, reporting only rare headaches and dizziness with exercise or emotional distress. We had no access to her MRI and did not know the degree of tonsillar herniation. She has a history of infertility and has undergone three previous uneventful procedures: one hysteroscopy two years ago, under general anesthesia with laryngeal mask, and two follicular punctions one year ago, under sedation maintaining spontaneous ventilation. She also refers a previous history of controlled asthma and penicillin allergy. Her preoperative evaluation was otherwise normal, and she was asymptomatic at the time of presentation. Due to fetal breech presentation and following neurosurgical advice, a cesarean delivery was planned.

A spinal block was performed at L3-L4 median approach with 10 mg hyperbaric bupivacaine and 0.001 mg sufentanyl through a G25 Quincke needle, by the most experienced anesthesiologist present. The spinal block was obtained with a T9 sensory level and hypotension ensued but was reverted with IV fluids (one litre of crystalloids infusion over thirty minutes) and a total of 30 mg ephedrine. At the extraction of the baby a total of 50 mg propofol was administered due to mild discomfort; a healthy 3240 g female newborn was delivered with an Apgar score of 10/10 at first/fifth minute. The remaining procedure was uneventful. Mother and baby were discharged home three days later. The patient´s sole complaint was neck stiffness that she devalued and attributed to insufficient sleep at the busy ward, and resolved after sleeping the first night at home. She denied headache or other neurologic symptoms. After a twenty month follow-up she remains minimally symptomatic without change of the intensity, frequency or pattern of previous symptoms.

We retrospectively gained access to her cerebral MRI, dated from 2007 (Fig.s 1-3), showing peg-shaped cerebellar tonsils displaced 8 mm below the level of the foramen magnum, with evidence of hindbrain overcrowding and obliteration of the subarachnoid space at the bulbo-medullar transition level. She has not undergone a full spinal MRI, so concomitant syringomyelia is unknown.

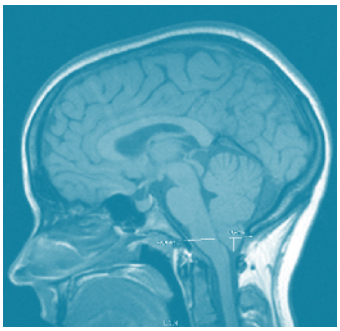

Figure 1 - Sagittal T1 MRI image showing peg-shaped cerebellar tonsils displaced 8mm below the level of the foramen magnum, with evidence of hindbrain overcrowding.

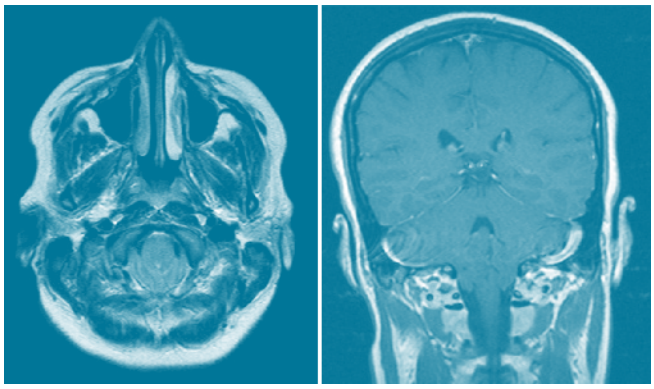

Figure 2 (Axial T2 MRI image) and 3 (coronal T1 MRI image with gadolinium): cerebellar tonsils displacement through the foramen magnum, resulting in obliteration of the subarachnoid space at the bulbo-medullar transition level.

DISCUSSION

CM-1 has an estimated prevalence of 0.1-0.5%. Patients might be asymptomatic or undiagnosed, with nonspecific complaints that are frequently devalued: in one case series of 364 symptomatic patients, at the time of definitive diagnosis, 59% had been told by at least one physician that their symptoms were psychogenic.[1] This might explain why literature is scarce and anesthetic management of CM-1 patients is still subject of debate. Both general and neuraxial anesthesia have safely been used for cesarean delivery in these patients,[3-6] but there are also reports of neurologic deterioration in patients with undiagnosed CM-1 submitted to spinal anesthesia[6] or after inadvertent dural puncture.[8-10] These symptoms may develop as late as two weeks after the dural puncture, with progressive worsening, probably due to slow cerebrospinal fluid leak with increasing craniospinal pressure gradient.[10]

General anesthesia is not without risks as laryngoscopy might increase intracranial pressure and also worsen the condition, and increased risk of pulmonary aspiration and hypoxia should be considered when deciding the anesthetic plan. Besides, keeping the patient awake enables constant neurologic monitoring anticipating a prompt intervention should any deterioration occur.

Concerning neuraxial anesthesia, a clean epidural block would certainly be expected to be safer than a spinal block, but in the event of an accidental dural puncture the outcome would probably be worse. There are several descriptions of epidural anesthesia, both for vaginal and cesarean delivery,[4,5] but we found only three previous reports of spinal anesthesia for cesarean delivery in uncorrected CM-1 patients, all described as uneventful.[4-6] Nevertheless, two of them were performed in the same patient and the degree of tonsillar herniation is not specified;[4] and the other report does not refer the follow-up time nor the degree of tonsillar herniation.[6] On the other hand, the sole report of neurologic deterioration after single-shot spinal anesthesia refers to a patient with severe CM-1 symptoms (cough related syncope, dysphagia

and obstructive sleep apnea), although undiagnosed.[7]

In our report, neck stiffness without headache developed after an uneventful spinal anesthesia. This could be related to worsening tonsillar herniation but is also a nonspecific symptom. In fact, it was a minor complaint and resolved spontaneously with proper rest after discharge from the hospital. Ideally, this patient should have had a recent cerebral MRI, moreover because she had been previously proposed for surgical treatment which she refused. Even so, at presentation she was asymptomatic, reporting rarely mild symptoms, which favours a current low degree of obstruction to cerebrospinal fluid circulation.

With this case, a total of four techniques in three patients have been described; more reports of positive outcome after spinal blocks in CM-1 patients are certainly needed before it is largely accepted as a safe technique. A multidisciplinary approach is needed and the anesthetic plan must be defined case by case, after a detailed evaluation that should include access to a recent MRI and neurosurgery consultation. Nevertheless, we believe spinal block might be a good option for uncorrected CM-1 patients, as long as they are only mildly symptomatic, without signs of elevated intracranial pressure, and that an extended follow-up period is guaranteed, due to possible late deterioration.

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