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CASE REPORT

The dilemma of choosing obstetrics and anesthesia techniques in a patient with cerebral cavernomatosis: a case report

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Abstract This report describes the case of a pregnant woman who arrived for preanesthetic assessment of External Cephalic Version (ECV) for fetus in breech presentation and cesarean section in case of ECV failure. Although the technique seems simple, attempts to rotate the fetus can result in elevated intracranial pressure, which might cause malformation bleeding. The most appropriate anesthetic technique in cases of arteriovenous malformations during C-sections has not been determined. Neuroaxial anesthesia is safe only in stable brain cavernomas, but the presence of spinal malformations contraindicates it. Anesthetic goals include stabilizing the blood pressure and reducing the risk of rupture.

Introduction

Cerebral Cavernomas (CC) are Arteriovenous Malformations (AVMs) with a prevalence of 0.1–0.5% in the general population.[1] Their most common symptoms are seizures (55%), focal neurological deficits (9%), nonspecific headaches (4%), and cerebral hemorrhages (32%).[1,2] Up to 25% of patients remain asymptomatic throughout their lives. Anesthetic and obstetric management of pregnant women with AVM is difficult as there are no clear guidelines.

Although External Cephalic Version (ECV) seems to be a simple technique, attempts to rotate the fetus increase the intra-abdominal pressure, resulting in an elevation of Intracranial Pressure (ICP) and cerebrospinal fluid pressure, which could be responsible for malformation bleeding. In these patients, it is also essential to maintain low intrathoracic and intra-abdominal pressures and avoid emesis. Since there is no literature about ECV in pregnant women with

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AVMs, it should be carefully performed by prioritizing maternal and fetal safety and hemodynamic stability.[2] If ECV fails, a Cesarean Section (CS) must be performed. Neuraxial Anesthesia (NA) is a safe choice for CS. However, the most appropriate anesthetic technique for cases of spinal or cerebral AVMs has not yet been determined. Anesthetic goals include stabilization of blood pressure and prevention of the risk of rupture.[1,3,4]

Although general anesthesia is a safe choice for pregnant patients with CC, NA can be used in stable brain cavernomas as it helps avoid the hemodynamic response associated with intubation and extubation and reduces the risk of aspiration. However, in cases of spinal cavernoma, NA should be avoided because of the risk of medullary ischemia. It is recommended to perform Magnetic Resonance Imaging (MRI) of the brain and spinal cord a year before pregnancy to guide anesthetic management. Cesarean delivery is not always required, and in cases of small lesions with no recent signs of bleeding, vaginal delivery can be performed. To our knowledge, this is the first case report of a pregnant woman with cerebral malformation undergoing an ECV.

Case report

We describe the case of a 41-year-old, 38-week pregnant woman, who arrived for preanesthetic assessment of ECV for fetus in breech presentation and CS in case of ECV failure. The patient was diagnosed with a left insular cavernoma with sporadic sensory crises in response to stress. The last MRI, performed one and a half a year before, revealed a stable cavernoma, and her neurologist's report stated that it had remained stable for years. Preoperative test results were normal.

The ECV was performed under sedation with 0.15 μg/kg/min of remifentanil. As it failed, a CS was performed under spinal anesthesia with a single puncture at the L3–L4 level and a 27G needle (10 mg of hyperbaric bupivacaine 0.5% and 20 μg of fentanyl), without any complications. Adequate uterine contraction was achieved by perfusion of 30 units of oxytocin and four misoprostol intrarectal tablets. The postoperative course was uneventful, and the patient was discharged on the third day.

Discussion

Cerebral cavernomas are AVMs detected on MRI in 0.1–0.5% of the general population,[1] accounting for 10–20% of all cases of AVMs. Up to 25% of patients remain asymptomatic throughout their lives. The most common symptoms of CC are seizures (55%), focal neurological deficits (9%), nonspecific headaches (4%), and cerebral hemorrhages (32%).[1] The highest number of cases are detected between the age group of 10 and 40 years (60–70%).[1] The familial incidence is approximately 20% and shows an autosomal dominant inheritance with variable expression and incomplete penetrance.[1] Familial malformations generally present with multiple lesions.

Genetically, CCs are associated with four loci: KRIT1 (CCM1) located on chromosome 7q11–22; MGC4607 (CCM2) on chromosome 7p13; PDCD10 (CCM3), originally identified as TF-1 cell apoptosis-related gene 15 (TFAR15), located on chromosome 3q26.1, and CCM 4 on chromosome 3q26.3–27.2.[1] These mutations alter the tight junctions between endothelial cells, resulting in gaps between them[1] and histologically produce hamartomatous vascular malformations with abnormally enlarged capillary cavities.

The fact whether pregnancy is a risk factor for hemorrhage from AVMs remains controversial.[4] Acute presentation of severe headache, meningism, and photophobia is characteristic of intracranial hemorrhage. The differential diagnosis should include eclampsia, arterial or venous intracranial thrombosis, tumors, abscesses, and inflammatory processes. Following hemorrhage, the maternal mortality rate significantly increases, and rupture of intracranial aneurysms or AVMs is responsible for 5–12% cases of maternal deaths.[4] Although AVMs do not cause hemorrhage, they can cause brain damage because of the ''steal phenomenon'', or by decreasing cerebral perfusion.[4]

In order to prevent hemorrhage, it is essential to avoid cranial hypertension in these patients, but ECV increases intra-abdominal pressure in the attempt to rotate the fetus, resulting in an elevation of ICP similar to the Valsalva maneuver, associated with dramatic changes in the venous pressure, cardiac output, and cerebrospinal fluid pressure, which can be responsible for malformation bleeding. In these patients, it is essential to maintain low intrathoracic and intra-abdominal pressures and avoid emesis. Therefore, the ECV should be carefully evaluated in these patients.

In the present case, ECV was performed under sedation with remifentanil at low doses. Remifentanil can be safely administered to pregnant women. It crosses the placenta and is rapidly metabolized and redistributed to both the mother and fetus, helps avoid hemodynamic response to pain and anxiety,[4] is an opioid with low emetic potential, and at low doses, the patient can maintain spontaneous breathing.

Vaginal delivery is not contraindicated in patients with small AVMs with no recent signs of bleeding, but large lesions or recent hemorrhages are relative contraindications for pregnancy and vaginal delivery.[1,3] An MRI of the brain and spinal cord performed in the previous year was needed to guide anesthetic management. In stable brain lesions, NA can be used, but the presence of spinal lesions contraindicates it.[1,3] In AVMs located in the brain, NA may be preferred because it avoids hemodynamic changes associated with general anesthesia. These changes should be controlled with nitroglycerin, remifentanil, lidocaine, and propofol during induction. Succinylcholine was contraindicated.[4] Invasive hemodynamic monitoring is recommended to maintain ICP and uteroplacental flow within appropriate limits. We should maintain low intrathoracic and intra-abdominal pressures[3] and avoid emesis associated with morphine and coughing during extubation.

Both the spinal and epidural blocks were used. Epidural blockage, being more gradual, seemed more suitable for hemodynamic stability, for decreasing nausea and vomiting, and avoiding the sudden increase in secondary ICP.[2] However, it can increase epidural pressure, thereby compromising the arterial blood flow and causing ischemic damage.[3] Spinal block can lead to hypotension, thereby decreasing blood flow to the brain; it also causes emesis. The use of vasoconstrictors (50–60% of patients) may also

promote spinal ischemia.[3] However, in the case of AVMs located in the spinal cord, NA is an absolute contraindication, although we do find exceptional cases describing its successful application.[3]

Spinal AVMs make the medulla more vulnerable to hypotension and ischemia. An arteriovenous fistula increases the intravascular pressure in the plexus responsible for spinal drainage, and the shunt decreases the collateral reserve in other areas, thus altering the venous drainage at the distal spinal segments, causing dilated epidural veins, and increasing the possibility of direct needle trauma.[3] Moreover, vasodilatation caused by the blockage may favor vascular theft in regions that are most sensitive to ischemia.

Finally, local anesthetics seem to decrease medullary blood flow, an additional reason to avoid NA in spinal cord AVMs. Oxytocin can be administered as a uterotonic, in small doses or infused because it does not seem to affect the cerebral blood flow, but methylergonovine, carboprost, and misoprostol should be avoided or administered with extreme caution, as they cause an increase in blood pressure and ICP.[5]

Patient perspective

The woman did not present any complication during any part of the procedure. She was happy for the preanesthetic assessment and for the intraoperative management. The patient gave her written consent.

Conclusion

To the best of our knowledge, this is the first clinical case report of ECV in a pregnant woman with CC. In our patient, it could only be performed owing to the stability of the lesion. The focal point of this case is to emphasize that the anesthetic choice is based on maternal and fetal safety and hemodynamic stability, thereby preventing the risk of rupture.[2] Both general and regional anesthesia have been used in these patients, and neither has been proven superior. In cases of spinal cavernoma, the literature aims to avoid NA. In all other cavernomas, NA is the first choice. However, since AVMs are rare, no definitive guidelines exist. It is recommended to perform brain and spinal MRI a year before pregnancy to guide anesthetic management. The management should be multidisciplinary, involving gynecologists, anesthetists, neurologists, and neurosurgeons.[4]

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Conflicts of interest

The authors declare no have conflicts of interest.

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