Cesarean Section in a Patient Diagnosed with Congenital Shortage of Coagulation Factors V and VII and an Arteriovenous Malformation Partially Embolized

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BACKGROUND

Arteriovenous malformations (AVMs) are congenital vascular abnormalities consisting of multiple arteries and veins connecting to each other as a fistula without the intervention of the capillary system¹. The prevalence of this condition is approximately 0.01 % to 0.5 % of the population². If symptoms are present, these occur before the age of 40. In females, symptom appearance concurs with their fertile age.

Inherited coagulation disorders are even rarer. The most common of them is factor VII deficiency. We present the case of a female patient suffering not only combined Factors V and VII deficiency but also a partially embolized intracranial AVM.

CASE REPORT

A 30-year-old primigravid patient with 39 weeks of gestational age was admitted to labor with intrauterine growth restriction. Patient was previously diagnosed with congenital deficit of coagulation factors V and VII and a partially embolized intracranial AVM.

After a multidisciplinary team discussion involving obstetrician, anesthesiologist and, hematologist, an elective cesarean section was performed. Recombinant human factor VII protein and multiple doses of tranexamic acid were administered before surgery. Rapid sequence induction was performed with propofol, rocuronium and a continuous infusion of remifentanyl. Remifentanyl infusion was continued for anesthesia maintenance among sevoflurane as Inhaled maintenance agent. No increased intraoperative blood loss was recorded and no blood products administration was required during surgery. Anesthesia emergence was uneventful and neurological postoperative assessment was normal. The patient and the newborn were discharged without any complications on the third postoperative day.

Figure 1. Cerebral Angiography: (A) Nidus arteriosus venous malformation. (B) Venous return.

DISCUSSION

Clinical manifestations of intracranial AVMs include seizures, headache and focal neurological deficit. Patients with AVMs might also present with intracranial hemorrhage. The long-term overall risk of hemorrhage has been evaluated in numerous studies at approximately 2%-3% of the patients diagnosed with intracranial AVMs per year.

According to the majority of the available published data, pregnancy is not a risk factor for hemorrhage from a brain with an AVM, yet this remains being very controversial¹. In the case presented, after thorough benefit-risk assessment, the decision was made to perform an elective cesarean section in order to avoid Valsalva maneuvers during labor. Besides that, our patient was diagnosed with factor V and VII deficiency, increasing the risk of epidural hematoma. Therefore, neuroaxial anesthesia was also avoided.

General anesthesia management of a patient with these conditions represents a great challenge. We should maintain hemodynamic stability during the whole case since hypotension might reduce uteroplacental blood perfusion and hypertension might increase bleeding risk from the cerebral AVM². Neonatal respiratory depression cases have been reported after remifentanyl administration; nevertheless, this respiratory depression might be transient. In our experience, remifentanyl should be considered as the opioid of choice during general anesthesia for its short duration of action and its potential benefit maintaining intraoperative hemodynamic stability.

CONCLUSION

Because of the limited published data about the management of AVM rupture during pregnancy and puerperium, the treatment for these patients is very difficult and controversial. Clinical decision must be made on an individual basis considering the risk and benefits of each conduct.

REFERENCES

1. Brown, R. D., Flemming K. D. et al. (2005). Natural History, Evaluation, and Management of Intracranial Vascular Malformations. *Mayo Clinic Proc.*
2. Lv X, Liu P., Li Y. (2015). The clinical characteristics and treatment of cerebral AVM in pregnancy. *The Neuroradiology Journal.*