CASE REPORT

Cerebral Arteriovenous Malformation in Pregnancy

Ma Saung Oo[1], Nik Rafiza Afendi[2], Mya Mya Thwin[3]

Abstract

Background: Cerebral arteriovenous malformations (CAVMs) are rare congenital lesions but the life-threatening condition with the possibility of rupturing during pregnancy.

Case description: The case was a 25-year-old primigravida at 17 weeks gestation who was presented with recurrent prolonged excessive vomiting for a 1-month duration and treated as hyperemesis gravidarum. Subsequently, she had right hemiparesis and it was preceded by throbbing headache. The clinical diagnosis of paradoxical emboli or ruptured CAVM is made based on the clinical findings. Magnetic resonance imaging (MRI) brain showed subacute bleeding of ruptured AVM at the left parietal region. Angiogram revealed the site of bleeding vessels for excision of AVM and helped to arrest the bleeding. The benefit of surgery was outweighing the risk of miscarriage and proceeded with craniotomy, evacuation of hematoma, and excision of AVM at 22 weeks of gestation. Her postoperative course was uneventful and there was no issue of teratogenicity as a fetus at 24 weeks gestation. She had delivered a baby boy 2.8 kg via elective cesarean section at 38 weeks gestation without maternal and fetal complications.

Conclusion: Cerebral arteriovenous malformations are relatively uncommon lesions in the general population and rare among pregnant females with significant maternal and fetal morbidity and mortality. The various clinical presentations warrant the different management approaches. Hence, a multidisciplinary team approach tailored to the individual case with appropriate management should be employed for pregnant women with CAVMs regardless of clinical presentation at gestational age and risk of rupture during pregnancy.

Keywords: Cerebral arteriovenous malformation, Hemorrhage, Multidisciplinary team, Pregnancy, Surgical management.

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Background

Cerebral arteriovenous malformation (CAVM) is a rare congenital lesion, but it is a life-threatening condition with a risk of rupture during pregnancy.[1] The appropriate timing and mode of delivery after intervention or observation of CAVM in pregnancy are ill-defined. One of the common symptoms of CAVM in pregnancy is prolonged refractory nausea and vomiting beyond the early second trimester.

Case Description

A 25-year-old lady, 17 weeks into her first pregnancy was presented with lethargy and giddiness due to recurrent nausea and vomiting for a 1-month duration. She had been admitted twice and treated as hyperemesis gravidarum for a similar problem. Transabdominal ultrasound showed a viable singleton fetus and all fetal parameters corresponded to the date. Blood investigation of full blood count and electrolytes results was normal except urine ketone was 2(+). Four days after admission, she had right-sided both upper and lower limbs weakness. It was preceded by a throbbing headache. However, she denied syncopal attack, seizure, and blurring of vision. Neurological examination revealed all cranial nerves were intact and right upper and lower limbs tone were normal but power 3/5 with plantar reflex noted. The early papilledema was noted on fundoscopy. The clinical diagnosis of paradoxical emboli or ruptured brain AVM was made based on the clinical findings. An echocardiogram revealed patent foramen ovale with left-to-right shunt and no endocarditis seen. An electroencephalogram (EEG) showed an abnormal slow wave of the left hemisphere.

A magnetic resonance imaging (MRI) brain revealed subacute bleeding of CAVM at the left parietal region (Fig. 1). Subsequently, an angiogram is performed to determine the site of bleeding vessels for excision of AVM and arrest the bleeding (Fig. 2). The decision was made by balancing the risks and benefits of surgical intervention. However, the benefit of surgery outweighed the maternal and fetal risk and proceeded with craniotomy, evacuation of hematoma, and excision of CAVM at 22 weeks of gestation. Intraoperative findings included 30 mL of blood clots near the ruptured vessels, which were evacuated, and the bleeding vessels were ligated before draining veins were excised and removed.

Subsequently, she was treated with Tegretol 200 mg twice per day and continued physiotherapy for the weakness of the right lower limbs. There was no teratogenicity issue as a fetus at 24 weeks gestation. She had delivered a baby boy 2.8 kg via elective cesarean section at 38 weeks gestation, and her postoperative course was uneventful. Both mother and baby are healthy and discharged home 10 days later.

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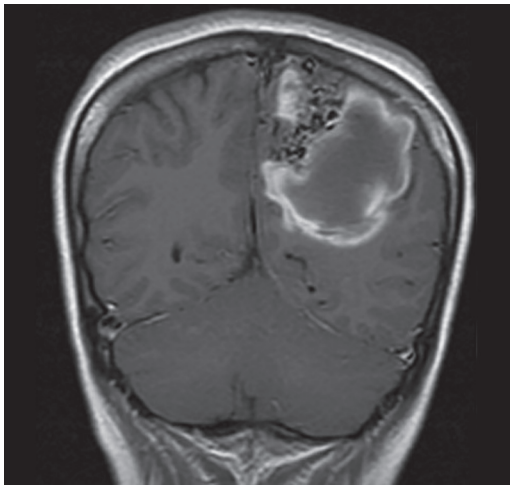

Fig. 1: Preoperative MRI image (coronal view) area of vascular malformation with adjacent hematoma in the left parieto-occipital region

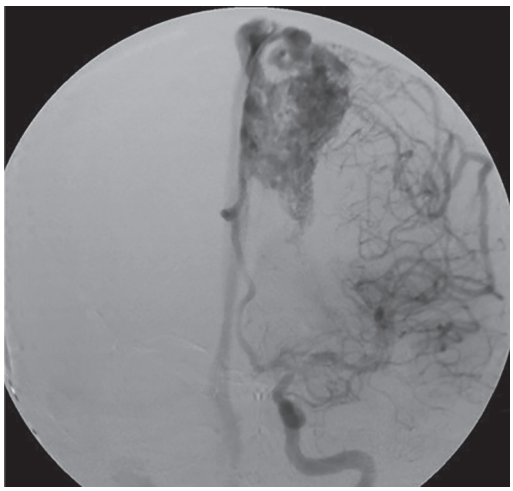

Fig. 2: Preoperative cerebral angiogram showed ruptured cerebral arteriovenous malformation

Discussion

Pregnancy coexisting CAVM is a challenging situation, and the natural history of CAVMs is poorly understood and even less understood in pregnant patients.[1] Pregnancy has been linked to increasing the risk of AVM rupture, and the physiological changes during pregnancy can complicate the AVM. It may be caused by the increased cardiac output or circulatory effects of elevated estrogen levels.[2]

Nausea and vomiting are one of the main symptoms in CAVM cases during pregnancy.[1] Hence, CAVM is the differential diagnosis for protracted prolonged nausea and vomiting in pregnant patients with associated neurological symptoms. The most common presenting sign of brain AVM is cerebral hemorrhage.[1] Most intracranial hemorrhages occur during the antepartum period. Nevertheless, history of previous AVM rupture is a known association with increased rebleeding from AVM, whether in the same pregnancy or later.[2] Ruptured CAVMs may affect the maternal and fetal outcomes during pregnancy.[2]

Until now, there is no consensus for definitive treatment guidelines of AVMs during pregnancy regarding timing and choice of appropriate intervention. Counseling these issues by explaining advantages and possible complications in detail based on the expert opinion was the adopted policy in current practice.[3] The risk-benefit counseling remains the gold standard for AVM intervention timing, except for emergency management of ruptured AVM, especially in clinically unstable patients. Therefore, the management of CAVM in pregnancy should be individualized after discussion with the patient and her family.

MRI and angiography are essential investigations to evaluate the abnormal vascular connection and determine the feeding vessels of AVM in pregnancy.[4] The benefits of angiography include better understanding the anatomy of the AVM with identification of feeding arteries, draining veins, and accessible feeders to facilitate surgical resection and minimize operative morbidity.[4] The fetal teratogenicity after 20 weeks gestation is minimal.

Open or endoscopic surgery with vascular resection or ligation, endovascular embolization, and stereotactic radiosurgery are the well-established management options for ruptured and unruptured cases on an individualized basis.[5] Urgent interventions such as emergent nidus resection, excision of abnormal vascular connection, ventricular drainage, and hematoma removal are reserved for patients with a ruptured AVM with or without neurological deficit.[3] Once rupture, timely emergency surgical intervention by excision of the AVM is crucial for good maternal and fetal outcomes, as observed in our case. Endoscopic microsurgery and adjuvant endovascular embolization in AVM in pregnancy are available options of choice.[5] Hence, temporary or definitive surgical intervention should be undertaken regardless of gestational age, and the pregnancy needs to progress despite everything concern for rupture AVM during pregnancy.

The determination of the time and mode of delivery depends on the joint multidisciplinary team's decision in pregnancy. The conservative management for an unruptured AVM in pregnancy appropriates if there is no increased risk of bleeding.

Due to the lack of clear guidelines, the explanation of timing, appropriate intervention, and associated risks and benefits with detailed planned procedure in advance is crucial for managing AVM in pregnancy. Elective cesarean is indicated if pregnancy is successfully continued for consideration of maternal and fetal benefits. When neurological deterioration occurs due to AVM rupture, emergency surgical intervention is necessary regardless of gestational age like our case. If the fetus is sufficiently mature, simultaneous cesarean section is possible.

Regarding the choice of the modality of intervention, we considered definitive surgical intervention in the ruptured case with the evacuation of hematoma and excision of AVM as one of the best options. The most difficult decision is the time and mode of delivery of unruptured pregnant cases with expectant treatment as the risk of bleeding during pregnancy is unpredictable. However, maternal and fetal risks of radiation or anesthesia are now minimal compares to the risk of rupture of AVM with either conclusive or conservative approaches. Hence, treatment should be individualized with consideration of other underlying associated risk factors during pregnancy.

Conclusion

Nausea and vomiting are typical symptoms during early pregnancy. Hence, a high index of suspicion should be made for the possibility of any other causes rather than hyperemesis gravidarum if the patient had a history of late-onset prolonged protracted vomiting.

Therefore, the management of AVM during pregnancy requires a multidisciplinary approach with close cooperation among the neurosurgeon, obstetrician, and anesthesiologist for successful maternal and fetal outcomes.

Clinical Significance

This article intended to bridge the gap between rare and common obstetrics and gynecology cases with a similar clinical presentation for definitive diagnosis with an appropriate management plan to achieve a favorable outcome.

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