Case report

Cerebral arteriovenous malformations during pregnancy: a management dilemma

Rubina Sohail,^{1,2} Qasim Bashir,³ Shahlla Kanwal,¹ Maryam Iqbal Ali²

¹Obstetrics & Gynaecology, Services Institute of Medical Sciences, Lahore, Pakistan
²Obstetrics & Gynaecology, Hameed Latif Hospital, Lahore, Pakistan

³Neurology, Hameed Latif Hospital, Lahore, Pakistan

Correspondence to
 Professor Rubina Sohail;
 rubina95@gmail.com

Accepted 25 September 2019

SUMMARY

The rupture of a brain cranial arteriovenous malformation (bAVM) has been associated with pregnancy; however, due to scarcity of data about this rare condition, management still remains a dilemma both for obstetricians and neurophysicians. The management plan is decided after weighing the benefits of bAVM treatment against the risk of bleeding in pregnancy. There is consensus on deciding the treatment of ruptured brain AVM during pregnancy based on neurological and not obstetrical indications. The management is decided using a multidisciplinary approach.

We report the case of a primigravida who presented at 15 weeks of gestation with intracranial haemorrhage secondary to ruptured bAVM, which was managed by glue embolisation. However, she presented again after 9 days in a moribund condition with fever, vomiting and malaise. The family was very concerned about the patient. The multidisciplinary team after evaluation of patient decided for hysterotomy based on the deteriorating maternal condition.

BACKGROUND

Cerebral arteriovenous malformation (AVM) is a rare congenital lesion that causes changes in the vasculature of the brain which increase possibility of rupture of these vessels, especially during pregnancy. The maternal physiological changes, like the increase in cardiac output and elevated oestrogen levels in pregnancy affect the structure of these vessels making them more prone to rupture.^{1,2} The rupture of an intracranial AVM in pregnancy is rare, but, it raises serious concerns for both maternal and fetal outcome. Surgical interventions for ruptured AVM during pregnancy would not only prevent episodes of rebleeding but also buy time till delivery. A close teamwork among neurosurgeons, obstetricians and anesthesiologists is required in managing this rare condition. It is essential to share all information, including treatment options, their possible side effects and fetomaternal outcome with the patient and the family. It is difficult to come up with an ideal treatment plan, as there are no definitive guidelines due to limited data and also because natural history of disease in pregnancy is not well understood.³⁻⁵

CASE PRESENTATION

A 20-year-old woman, at a gestation of 15 weeks, presented to the hospital emergency with history of left sided hemiparesis (weakness) for 18 days.

Her medical history was insignificant. On physical examination, there was left sided hemiparesis and Glasgow coma scale (GCS) at presentation was 12/15.

INVESTIGATIONS

Haemoglobin was 10 g/L, random blood sugar 77 mg/dL, liver function and kidney function tests were normal.

Non-contrast CT brain scan at the transferring institution demonstrated acute right basal ganglia intracranial haemorrhage (ICH) and hydrocephalus. MRI revealed right ICH extending to the basal ganglia, thalamus and frontal lobe with flow voids suggestive of underlying AVM. Because of mass effect, right lateral ventricular compression and significant hydrocephalus were present. Non-contrast CT brain was repeated and it revealed a heterogeneous rather dense mass (50×55 mm AP×TR) (AP, anteroposterior; TR: repetition time) in right deep parietal region showing foci of calcification. Mass showed haemorrhage with intraventricular extension of bleed. There was significant surrounding peri-lesion oedema. Lesion showed mass effect with significant midline shift (video 1).

Obstetrical ultrasonography showed a single fetus with a biometry of 16.2 weeks with anterior upper midsegment placenta. There were no obvious fetal anomalies. Fetal heart rate was 167 beats/min.

Urine report on second admission showed field full of pus cells. On urine culture, growth of *Escherichia coli* was identified.

TREATMENT

The patient was pregnant with a ruptured bAVM. Therefore a multidisciplinary team consisting of obstetrician, neurosurgeon and intensivist assessed the patient and counselled the family extensively regarding the treatment options. A joint consensus was reached to reduce the size of AVM nidus via glue embolisation to facilitate her recovery by reducing the AVM induced mass effect. There was a 5% risk of re-bleed in case of incomplete nidus obliteration, which was explained to the family. Intravenous mannitol therapy was initiated for hydrocephalus.

The procedure was performed under general anaesthesia. Lead aprons to protect the fetus from radiation covered the patients' abdomen and pelvic areas. A 6 Fr short sheath was placed in the right femoral artery. Then, a 5 Fr cerebral VI guide catheter over Terumo 0.035 mm glide wire was used to perform the diagnostic digital subtraction

© BMJ Publishing Group Limited 2019. No commercial re-use. See rights and permissions. Published by BMJ.

To cite: Sohail R, Bashir Q, Kanwal S, et al. *BMJ Case Rep* 2019;**12**:e228759. doi:10.1136/bcr-2018-228759

Video 1 Video of CT Scan.

angiography (DSA) cerebral angiography. A Spetzler Martin Grade III right hemisphere AVM was visualised with arterial feeders originating from right middle cerebral artery (MCA) branches. A giant venous aneurysm measuring approximately 4 cm was also visualised. Right A1-anterior cerebral artery (ACA) was noted to be congenitally absent and right ACA territory filling seen via left anterior communicating artery. Angiographically vascular stealing from right MCA into the AVM was demonstrated with distal right MCA territory being retrogradely via the right ACA and right posterior cerebral artery branches.

Then, a 6Fr Chaperone guide catheter over Terumo glide wire was advanced and positioned in the right distal cervical internal carotid artery (ICA). Patient was systemically heparinized, and serial activated clotting time (ACTs) were obtained to achieve ACT >250–300 s.

Following this, a Headway Duo Micro catheter (Microvention, USA) over Traxcess Microwire (Microvention) was advanced, and a single large arterial pedicle was glue embolized using 20% diluted Glubran glue. Post glue embolisation angiography demonstrated noticeable reduction in the AVM filling and giant venous aneurysm filling. Postprocedure, she was

Video 2 Video of Glue Embolization.

transferred to intensive care unit (ICU) for postprocedure care ([figure 1](#), [video 2](#)).

After procedure, her overall recovery was slow, and she persisted with complaints of nausea, vomiting and poor oral intake. She was discharged after a couple of days after stabilisation. At home, her symptoms reverted and she had to be readmitted 9 days later with fever of 38.3° C and disorientation. A diagnosis of urinary tract infection was made based on the urine report. There was no other nidus of infection. CT brain done at the time showed only a mild reduction in the severity of hydrocephalus. The family was extremely concerned about the deteriorating clinical condition of the patient. The family was counselled about the slow recovery process because of residual mass effect from initial haemorrhage in pregnant patients. The multidisciplinary team after consulting the family decided for termination of pregnancy by hysterotomy. Meanwhile, she was given intravenous antibiotics, steroid, antiepileptic, antiemetics and mannitol along with supportive management by the ICU staff. Her postoperative recovery was satisfactory. She became well oriented, and fever settled. She, however, had residual weakness of her left arm and leg at the time of discharge.

OUTCOME AND FOLLOW-UP

At 1 month post hospital discharge, neurology follow-up was found to be ambulatory with minimal assistance. Speech and

Figure 1 Glue embolisation. (A) Right ICA AP view—arterial phase. Note: the AVM nidus and early draining vein and associated giant venous aneurysm filling. (B) Right ICA lateral view—venous phase showing giant venous aneurysm. (C) Right ICA angiography (AP view) Post glue embolisation. Note: angiographically visible reduction in AVM nidus size and delayed filling of the draining vein aneurysm. AVM, arteriovenous malformation; AP, anteroposterior.

comprehension functions were completely intact, and left arm proximal motor strength was graded at 3/5 and hand 2/5 with spasticity. She was recommended to undergo repeated MRI brain study at 3 months and continue with out-patient physiotherapy till then.

DISCUSSION

Cerebral AVM is a rare congenital lesion (0.01%–0.5%), which is an abnormal communication between cerebral arterial and venous systems with no intervening capillary bed.^{1–3} This affects women in their third to fifth decade of life and usually presents before 40 years of age.^{3 5–7} Cerebral AVM has a high possibility of rupture during pregnancy due to increase in cardiac output and elevated oestrogen levels in pregnancy.^{1–3 5 6} However, the majority of large epidemiological studies of intracerebral haemorrhage in pregnancy have not been differentiated by mechanism. Risk of re-bleeding in the same pregnancy is 27%–30% which is much higher than women who are not pregnant within a year.^{1 3 4 8 9}

Literature has reported an increased risk of cerebral infarction and intracerebral haemorrhage during puerperium (6 weeks after delivery).^{10–14} Though the rupture of an intracranial AVM in pregnancy is rare (0.6%–3.5%), however, it leads to serious fetomaternal complications.¹⁵ Regarding maternal mortality due to AVM, there are multiple studies quoting an incidence of 10%–40%^{6 7 15–17} and fetal mortality of 14%.⁷

In this patient surgical intervention was decided by the multidisciplinary team to prevent episodes of re-bleeding and to buy time till delivery.

Different treatment strategies have been reported in literature to manage ruptured AVM in pregnancy. Ruptured AVM has been successfully being treated by surgical resection.^{3 15} Direct open surgery is considered to be associated with a lower rate of fetomaternal mortality when compared with conservative management alone, however, it is associated with an elevated risk of intraoperative bleeding which causes a decrease in uterine and placental blood supply and hence risk to fetus.⁴ Non-surgical options like endovascular embolisation have shown promising results in many case reports.^{1 4 15}

Studies have reported successful management when embolisation is followed with surgical resection.¹⁵ Kalani *et al* concluded after studying 64 cases of cerebral cavernous malformations during pregnancy that this condition is not a contraindication to pregnancy or vaginal delivery, although some earlier conditions had suggested that.^{16 17}

Although, there is no evidence of vaginal delivery being associated with increased risk of bleeding. However, there is divided opinion on the ideal mode of delivery. Caesarean section maybe safer as it will enable better control of maternal blood pressure and avoid haemodynamic changes of intravascular pressure preventing stress on vessel walls.⁷ Mode of delivery after complete removal of AVM can be decided on obstetrical grounds.

In our patient, glue embolisation was done in the hope that the condition of the patient would improve, and termination of pregnancy was not considered as an option at the time. However, when the patient presented again with complications and fever, the multidisciplinary team were worried about the deteriorating condition of the patient and possibility of re-bleed

was considered. Hence, the pregnancy was terminated by hysterotomy after consultation with the family.

Learning points

- ▶ There is a consensus on deciding the treatment of a ruptured brain arteriovenous malformation during pregnancy based on neurological and not obstetrical indications.
- ▶ The management of patients with this rare condition should be decided using a multidisciplinary approach with close collaboration of obstetricians.
- ▶ Targeted glue embolisation may be an effective management in selected cases where flow reduction might have a protective effect.

Twitter Rubina Sohail @rubinasohail@rubinasohail

Contributors RS, SK: manuscript writing; MIA, QB: management and manuscript writing.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Schalet G, Shah M. Endovascular management of cerebral arteriovenous malformations in pregnancy: two case reports and a review of the literature. *J Neurol Res* 2012;2:215–20.
- 2 Trivedi RA, Kirkpatrick PJ. Arteriovenous malformations of the cerebral circulation that rupture in pregnancy. *J Obstet Gynaecol* 2003;23:484–9.
- 3 Rispoli R, Donati L, Bartolini N, *et al*. Rupture of an intracranial arteriovenous malformation (AVM) in pregnancy: case report. *J Stem Cell Res Ther* 2015;5:2.
- 4 Asano S, Hayashi N, Edakubo S, *et al*. Successful perinatal management of a ruptured brain arteriovenous malformation in a pregnant patient by endovascular embolization followed by elective cesarean section: a single-case experience. *JA Clinical Reports* 2016;2.
- 5 Mohamed A, Ahmed S, Tamer H, *et al*. Cranial arteriovenous malformations during pregnancy: a multidisciplinary algorithm for safe management. Case series and review of the literature. *J Neurol Stroke* 2016;4.
- 6 Lv X, Liu P, Li Y. Pre-Existing, incidental and hemorrhagic AVMs in pregnancy and postpartum: gestational age, morbidity and mortality, management and risk to the fetus. *Interventional Neuroradiology* 2016;22:206–11. 10.1177/1591019915622161
- 7 Lv X, Liu P, Li Y. The clinical characteristics and treatment of cerebral AVM in pregnancy. *Neuroradiol J* 2015;28:234–7.
- 8 Wilkins RH. Natural history of intracranial vascular malformations: a review. *Neurosurgery* 1985;16:421–30.
- 9 Uchide K, Terada S, Akasofu K, *et al*. Cerebral arteriovenous malformations in a pregnancy with twins. *Neurosurgery* 1992;31:780–2.
- 10 Gross BA, Du R. Hemorrhage from arteriovenous malformations during pregnancy. *Neurosurgery* 2012;71:349–56.
- 11 Porras JL, Yang W, Philadelphia E, *et al*. Hemorrhage risk of brain arteriovenous malformations during pregnancy and puerperium in a North American cohort. *Stroke* 2017;48:1507–13.
- 12 Kittner SJ, Stern BJ, Feeseer BR, *et al*. Pregnancy and the risk of stroke. *New England Journal of Medicine* 1996;335:768–74.
- 13 Ros HS, Lichtenstein P, Bellocco R, *et al*. Increased risks of circulatory diseases in late pregnancy and puerperium. *Epidemiology* 2001;1:456–60.
- 14 Liu X-j, Wang S, Zhao Y-l, *et al*. Risk of cerebral arteriovenous malformation rupture during pregnancy and puerperium. *Neurology* 2014;82:1798–803.
- 15 Jermakowicz WJ, Tomycz LD, Ghiassi M, *et al*. Use of endovascular embolization to treat a ruptured arteriovenous malformation in a pregnant woman: a case report. *J Med Case Rep* 2012;6:113.
- 16 Dias MS, Sekhar LN. Intracranial hemorrhage from aneurysms and arteriovenous malformations during pregnancy and the puerperium. *Neurosurgery* 1990;27:855–66.
- 17 Kalani MYS, Zabramski JM. Risk for symptomatic hemorrhage of cerebral cavernous malformations during pregnancy. *J Neurosurg* 2013;118:50–5.

Copyright 2019 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <https://www.bmj.com/company/products-services/rights-and-licensing/permissions/>
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

Customer Service

If you have any further queries about your subscription, please contact our customer services team on +44 (0) 207111 1105 or via email at support@bmj.com.

Visit casereports.bmj.com for more articles like this and to become a Fellow