Maternal arteriovenous malformation in pregnancy from presentation to birth: case report and literature review

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ABSTRACT

We describe the case of 37-year-old, gravida 9 para 4, she was 13 weeks and 4 days pregnant who had ruptured cerebral aneurysm adjacent to large frontal arteriovenous malformation causing intracranial haemorrhage. Underwent urgent craniotomy and repair. Medical and surgical therapy to maintain cerebral pressure. Multidisciplinary team meeting arranged for combined maternal and fetal wellbeing. Patient developed recurrent episodes of rising temperature and tachycardia with chorioamnionitis out-ruled. Final multidisciplinary meeting occurred at 31 weeks of pregnancy advised for elective cesarean section under general anaesthesia at 34 weeks, given ongoing urinary tack sepsis. Cesarean section performed, healthy 2.6 kg baby girl delivered, Minimal neonatal resuscitation required. This case showed good maternal and fetal outcome achieved as patient's modified Rankin scale(mRS) (score measures the degree of disability or dependence in the daily activities of people who have suffered a stroke or other causes of neurological disability) improved from 5 to 4 and complete resolution of sepsis signs including tachycardia. Cooperation between neurosurgeons, obstetricians, neonatologist and anaesthesiologists and treatment strategy given to the patients are essential, guidelines for pregnant patients with cerebral arteriovenous malformation should be established.

KEYWORDS

Arteriovenous malformation, intracranial haemorrhage, chorioamnionitis, maternal, fetal medicine.

Background

The prevalence of cerebral arteriovenous malformation is approximately 0.01%–0.05% in the general population [1]. It affects men and women equally and commonly presents in patients aged 20–40 years old. Arteriovenous malformations a complex vascular mass or nidus that shunt blood between arterial-venous circulations without an intervening capillary bed. Although patients are usually asymptomatic, they are at increased risk of rupture. This results in management dilemma in clinical practice [1]. In the United Kingdom and Republic of Ireland in 2013–15 there were 12 women who died from intracranial haemorrhage during or up to six weeks after pregnancy, 7 from subarachnoid haemorrhage and 5 from intra cerebral haemorrhage. This represents an overall maternal mortality rate directly due to intracranial haemorrhage of 0.48 per 100,000 maternities [2]. In a prospective and population-based study, researchers found that pregnancy did not increase arteriovenous malformation bleed rates, but risk of intracranial hemorrhage was increased during the postpartum period [3].However recent Study in Japan in 979 patients over period of 50 years concluded No increased risk of hemorrhage was found in patients with cerebral arteriovenous malformation during pregnancy and the puerperium [4].

Physiologically known that increase in cardiac output during pregnancy and intrapartum may cause arteriovenous malformation to rupture. Majority of cases with arteriovenous malformation rupture happens between 18 to 20 weeks of gestation and within post-partum period. However, there are contradictory results among investigators regarding the risk of arteriovenous malformation rupture during pregnancy. Meanwhile, Horton *et al*. found that risk of arteriovenous malformation bleed during pregnancy was 3.5%, similar to the general population (3.1%) [5].

The nature of arteriovenous malformations is poorly understood, and even less understood in pregnant patients, because the frequency is rare. No definitive guidelines for the treatment of arteriovenous malformations during pregnancy exist and the management of cerebrovascular disease in pregnancy is under discussion [6-9]. A case series of 9 pregnant women with arteriovenous malformation six patients presented with intra-cerebral hemorrhage during pregnancy (first hemorrhagic episode), 3 of 6 patients who presented with intra-cerebral hemorrhage, arteriovenous malformation removal was performed during pregnancy. One patient required emergency surgery for the mass effect of the hematoma, and 2 patients with Spetzler-Martin grade I and II (This arteriovenous malformation grading sys-

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tem allocates points for various features of intracranial arteriovenous malformation to give a grade between 1 and 5. Grade 6 is used to describe inoperable lesions, the score correlates with operative outcome) arteriovenous malformations underwent elective surgery for the prevention of re-bleeding. intra-cerebral hemorrhages occurred in the 21st week, 16th week, and 25th week of gestation, the management of pregnancy after removal of the arteriovenous malformation was similar to a normal pregnancy, with vaginal delivery in one case and cesarean section in two cases respectively, patients modified Rankin scale (mRS) score was 0 [10]. Anesthetic management of pregnant women with ruptured arteriovenous malformation poses multiple challenges to the anesthesiologist. Anesthetic techniques during cesarean section in these patients should ensure precise hemodynamic control [11].

What makes this case unusual are its early presentation, multiparity of the patient, severity of the intra-cerebral hemorrhage with modified Rankin scale (mRS) Score of 5 and continuation of pregnancy to third trimester for delivery via cesarean section.

Case presentation

37 years gravida 9 para 4, she was 13 weeks and 4 days pregnant presented to a hospital in France following sudden onset severe headache, associated with photophobia, nausea and vomiting. The Glasgow Coma Scale/Score (GCS) (estimates coma severity) at presentation was 9/15, this deteriorated overnight to 6/15, requiring intubation and ventilation. Non-contrast computerized tomography (CT) brain showed intracranial and intra-ventricular haemorrhage secondary to ruptured cerebral aneurysm adjacent to large frontal arteriovenous malformation (Figure 1). Patient underwent urgent craniotomy and arteriovenous malformation repair. In intensive care unit (ICU) received medical therapy to control increased intracranial pressure (ICP) to maintain it less than 20mmHg. An external ventricular drain was inserted. External ventricular drain migrated to fourth ventricle prior to removal. There was no associated haemorrhage. Nimodipine and leviteracetam were given for vasospasm and seizure prophylaxis respectively. Percutaneous tracheostomy was inserted to maintain airway patency. Additionally, treatment was administered for three ventilator acquired pneumonias and urinary track infection.

History of current pregnancy was unremarkable prior to cerebral haemorrhage. This was a planned pregnancy, presented for booking at 8 weeks, with all booking bloods normal. Past medical and surgical history revealed nil of note. Past obstetrics history was significant for nine prior pregnancies; four carried to term and delivered vaginally and four prior first trimester miscarriages. Two latter pregnancies were complicated by gestational diabetes, requiring induction of labour at 38 weeks and 6 days of gestation.

At 18 weeks and 4 days gestation, she repatriated to Beaumont Hospital (the national neurosurgical centre). Magnetic resonance imaging (MRI) brain prior to transfer showed evidence of persistent mild ventricular dilatation and frontal lobe infarct with GCS of 6/15 (figure 2, 3). In ICU management coordinated between neurosurgery, anaesthetics and gynaecology

Figure 1 CT brain showed intracranial and intraventricular haemorrhage.

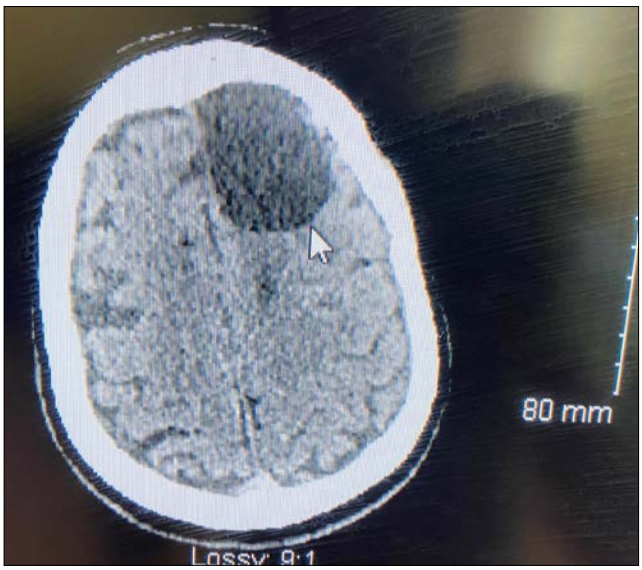


Figure 2 MRI: frontal lobe infarct.

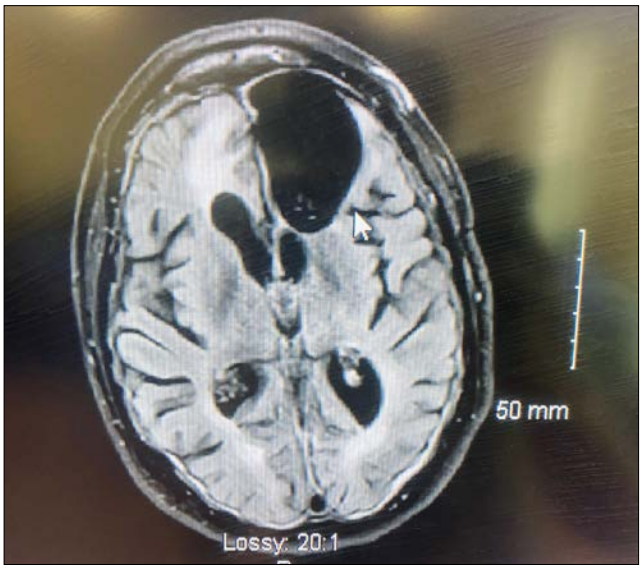
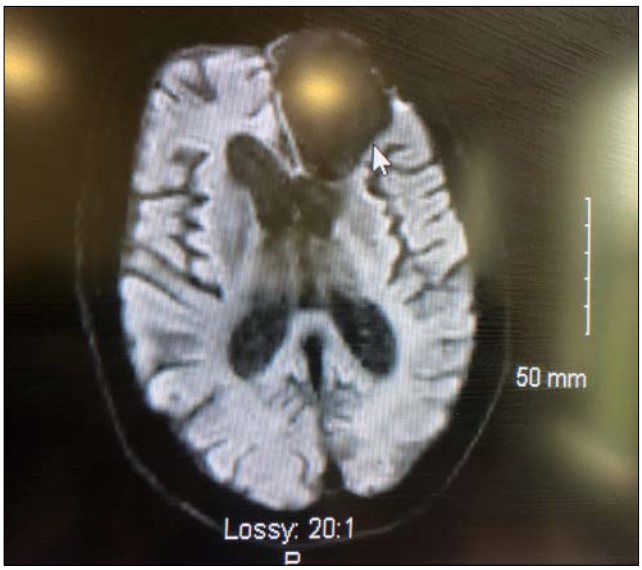


Figure 3 MRI: persistent mild ventricular dilatation.



team. As patient was stable from neurosurgical perspective, no further intervention was required. Extensive MDT rehabilitation was initiated with input from physiotherapy and occupational therapy to preserve functionality, prevent contractures and dieticians to initiate nasogastric feeds. Obstetrics plan was twice weekly fetal heart rate monitoring, plan for fetal anomaly scan at 21weeks of pregnancy fortnightly MDM discussions with fetal growth scans, commencement of thromboprophylaxis, administration of antenatal steroids at 24 weeks gestation and oral glucose tolerance test at 24-26 weeks of pregnancy to assess for gestational diabetes with aim for term delivery at 37 weeks gestation. Fetal anomaly ultrasound performed at 24weeks, which showed normal fetal anatomy and Estimated fetal weight 1000g, which was equivalent to date. Was transferred for management at ward level at 26 weeks of pregnancy.

Subsequent MDT discussions opted to keep tracheostomy weaning due to increased intra-abdominal pressure and difficulty with ventilation. Botox injections were initiated to ease contractures of upper limbs. Endocrine input was sought, with oral glucose tolerance test confirming gestational diabetes at 26 weeks. This was initially managed with diet control however; fetal growth chart was at 85th centile, basal Levemir was commenced and titrated accordingly over course of pregnancy. HbA1c (measure of how well controlled your blood sugar has been over a period of about 3 months normal level is lower than 48 mmol/L) remained normal at 32mmol/L. Blood glucose targets were elevated (from normal gestational diabetes levels) due to presence of acquired brain injury. Haematology input was also sought regarding anaemia, with Hb of 9.6g/dL. This was deemed multifactorial in nature and likely secondary to physiological haemodilution, recurrent phlebotomy and systemic stress response. An iron infusion was given, with Hb rising to 10.4g/dL.

Condition complicated by recurrent episodes of pyrexia, first episode was recorded at 21 weeks. Septic screen showed raised inflammatory markers. Chorioamnionitis was out-ruled by normal abdominal exam and no vaginal discharge, in addition to no clinical evidence of fetal distress on Ultrasound. Tracheal aspirates and Midstream urine samples were taken, both were positive for pseudomonas. Several courses of antimicrobials were completed including meropenam, vancomycin and pipercillin-tazobactam on advice from microbiology. After elective re-admission to high dependency unit at 30 weeks given level of observation required, with Persistent tachycardia remained an issue with baseline heart rate 140-150 beat per minute, Cardiology advised commencement of 1.25mg bisoprolol, with aim of heart rate less than 120 beat per minute.

Final MDM occurred at 31 weeks. Maternal fetal medicine colleagues advised for elective cesarean section under general anaesthesia at 34 weeks, given ongoing urinary track sepsis with cultures positive for multidrug resistant pseudomonas. Additionally, there was evidence of polyhydramnios and unstable lie, with fetus in breech position on ultrasound at 31 weeks secondary to gestational diabetes. Due to fetal risk of hypoglycaemia and spontaneous breech labour. Received both doses of dexamethasone 12mg 24 hours prior to delivery. Thromboprophylaxis held 12 hours prior to the operation. Antimicrobial prophylaxis in form of daptomycin & gentamicin was administered on induction of anaesthesia.

Cesarean section performed at 34 weeks under general anaesthesia. Neonatology and midwifery were in attendance. Operative course was uncomplicated with healthy 2.6 kg baby girl delivered, cried spontaneously. Minimal neonatal resuscitation required, with skin to skin initiated. Estimated blood loss intra-operatively was 1.5L. There were no acute maternal concerns postoperatively.

In high dependency unit, tachycardia settled to 90 beat per minute and no further episodes of pyrexia. Lactation was suppressed using cabergoline 1.25 mg once a day for 14 days. Oral glucose tolerance test was be performed 6 weeks postpartum to assess resolution of gestational diabetes. From neurological perspective, maternal condition has improved, with GCS 9/15 and visual fixation to voice. Initial stages of tracheostomy weaning have commenced. Extensive MDT rehab continues, with plan for transfer to dedicated rehabilitation facility when tracheostomy weaning completed. Long term prognosis from neurological perspective remains guarded.

Discussion

AVM is generally present in patients aged between 20 and 40 years, and is more common in those over 30 years, the childbearing age for women [12].

Maternal management of patients with ruptured AVMs should be based mainly on neurosurgical indications rather than on obstetrical indications. When neurological deterioration occurs due to AVM rupture, emergency surgery is necessary. If the fetus is sufficiently mature, simultaneous cesarean section is possible. When there is no indication for emergency surgery for AVM, blood pressure management is important [13]. Our patient underwent craniotomy and AVM repair then medical therapy to control increased intracranial pressure. An external ventricular drain was inserted.

In addition to the maternal and neurosurgical treatment priorities, consideration of the fetus is also necessary and cooperation between obstetricians and anesthesiologists is essential during surgery. If the fetus has reached the minimum age for extrauterine life, obstetricians prepare for emergency cesarean section in case of fetal distress. Extensive MDT rehabilitation was initiated with input from physiotherapy and occupational therapy. Initial obstetrics plan was; twice weekly fetal heart rate monitoring, plan for fetal anomaly scan at 21 weeks of getation, fortnightly MDM discussions with fetal growth scans, commencement of thromboprophylaxis, administration of antenatal steroids at 24 weeks and oral glucose tolerance test at 24-26 weeks to assess for gestational diabetes with aim for term delivery at 37 weeks. Subsequent MDT discussion opted to start botox injections to ease contractures. Endocrine input was sought, with OGTT confirming GDM at 26 weeks, initially managed with diet control then basal Levemir was commenced. Final MDM occurred at 31 weeks advised for elective cesarean section under general anaesthesia at 34 weeks given ongoing urinary track sepsis which treated with IV ciprofloxacin as cultures positive for multidrug resistant pseudomonas. tachycardia was treated with 1.25mg bisoprolol.

If the AVM is completely resected during pregnancy, the method of delivery can be determined based on the obstetrical indications [7]. In patients with AVM during pregnancy, problems during labor are related to the excessive cerebral hemodynamic changes, and cesarean section tends to be performed in these circumstances [11].

Conclusions

We achieved good maternal and fetal outcome in our case as patient's modified Rankin scale improved from 5 to 4 and complete resolution of sepsis signs including tachycardia. Cooperation between neurosurgeons, obstetricians, neonatologist and anesthesiologists, and sufficient information about the treatment strategy given to the patient are essential. Finally, for better maternal and fetal prognosis, guidelines for pregnant patients with cerebral AVMs should be established.

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