Anaesthetic management of vein of Galen malformation in a very low birth weight preterm baby for endovascular embolisation

Rajani MP Ramakrishnan, Shwetal U. Goraksha, Bhoomika P. Thakore, Joseph N. Monteiro, Manju T. Butani

Abstract

Extra-cardiac arteriovenous malformations are rare causes of severe cardiac failure in the neonatal period. Aneurysmal malformation of the vein of Galen may lead to a diagnostic confusion because the presenting signs point to a cardiac cause of failure. Endovascular therapy is found to be an effective and safe therapeutic modality available to treat these lesions. Intervention in the newborn is difficult and is preferably postponed until about 5 months. We present the anaesthetic management of a very low birth weight preterm neonate with a vein of Galen malformation who needed intervention within the first week of life.

Key words: Endovascular embolisation, preterm, vein of Galen malformation

INTRODUCTION

Vein of Galen malformations (VOGMs) are rare anomalies of the intracranial circulation that constitutes 1% of all intracranial malformations. However, they represent 30% of vascular malformations presenting in the paediatric age group.[1,2]

The nomenclature is imprecise because the dilated venous structure that is characteristic of this malformation represents the embryonic median prosencephalic vein and not the vein of Galen itself. Patients usually present with neurological manifestations and can present with severe cardiac failure in the neonatal period.

Management of these lesions by definitive intervention is challenging, especially in the neonatal period.

Endovascular therapy has been found to be a safe and effective therapeutic modality to treat these lesions. Intervention in the newly born is difficult and is usually postponed until about 5 months of age, balancing the benefits of safe embolisation against the risk of cerebral damage.[3]

We report the anaesthesia management of a neonate with VOGM who needed emergency therapeutic intervention within the first week of life.

CASE REPORT

A preterm neonate delivered by caesarean section for foetal distress, with a very low birth weight of 1.75 kg, developed respiratory and severe cardiac failure soon after birth. On systemic examination, the child...
Ramakrishnan, et al.: Anaesthetic management of vein of Galen malformation in a very low birth weight preterm baby for endovascular embolisation

had a hyperdynamic precordium with a pansystolic murmur in the mitral area and hepatomegaly. The blood investigations revealed a deranged coagulation profile with a raised activated partial thromboplastin time - 51.9 seconds. Serum electrolytes were Na - 140 meq/l, K - 4.2 meq/l and the arterial blood gas showed PH - 7.3, PCO₂ - 43.5, PO₂ - 70.5, HCO₃ - 21.8 on FiO₂ 0.4. Two-dimensional echocardiography demonstrated severe pulmonary arterial hypertension with a ventricular septal defect with the right to left shunt. A prenatal ultrasonography had suggested a VOGM and a magnetic resonance angiogram confirmed a large VOGM directed towards the left [Figure 1].

Cardiac failure was treated with digoxin and enalapril. However, it was unsuccessful in controlling the high output failure. The Bicetre neonatal evaluation score was used to monitor progress daily. When the score dropped from 15 to 11 on day 4, an emergency endovascular embolisation was planned.

After confirming consent and adequate starvation, all standard monitors such as electrocardiography, ETCO₂ pulse oximeter, non-invasive blood pressure cuff and temperature probe were attached. Intra-operatively the heart rate was maintained at 120–130/min, saturation was 98–99%, ETCO₂ at 30–32 mmHg and PaCO₂ at 36–39 mmHg. Radiant warmers were used, and patient was adequately wrapped. The patient had a 24G intravenous catheter with a titrated dopamine infusion on flow. The child was induced with sevoflurane using JR-circuit and after confirming ventilation, muscle relaxant atracurium 0.5 mg/kg was given, and the patient was intubated with a 3.0 mm uncuffed endotracheal tube. Anaesthesia was maintained with sevoflurane, air: oxygen and fentanyl. The femoral artery was cannulated with a 5 French sheath and the sidearm of the sheath was transduced to monitor arterial pressure intra-operatively. The femoral vein was also secured. Since the patient was already on the inotropic support, we maintained normotension throughout the procedure. N-butyl-cyanoacrylate glue was used to embolise the feeding vessels - 60% obliteration was achieved [Figures 2-4].

The procedure lasted for 4 h. The baby received crystalloids, 0.45% DNS with KCl, at 6 ml/h throughout the procedure. Since there was blood loss through the femoral sheath, we also gave a total of 30 ml of packed red cells and 30 ml of fresh frozen plasma to correct the loss. At the end of the procedure, the child had hypothermia and was shifted to the Neonatal Intensive Care Unit for elective ventilation and for further management. The baby’s cardiac function improved almost immediately after the intervention. He started responding to the cardiac failure treatment. He was extubated on the 4th day after the procedure. The baby also underwent an uneventful bilateral inguinal hernia repair 3 weeks later under general anaesthesia and was later discharged from the hospital a week after the surgery. After 6 months, a second stage embolisation was performed.

DISCUSSION

VOGMs are a rare and diverse group with a complex anatomy, pathophysiology and serious clinical sequelae. VOGM is a misnomer. It is in fact the median vein of the prosencephalon, the embryonic precursor of the vein of Galen. These malformations are congenital vascular malformations made from multiple arterial feeders establishing direct or indirect shunts with large median vein collector. The latter is not the vein of Galen but a persistent embryonic channel that normally participates information of vein of Galen.[3,4] This usually occurs between 6th and 11th week of intrauterine life.[1,5]
Patients present with cardiac and neurological manifestations depending upon the age of presentation. Neonatal manifestations can range from asymptomatic cardiomegaly to severe cardiac failure that is refractory to medical management. Infants and children usually present with macrocephaly or with hydrocephalus. Cardiac manifestations are absent or very mild. Patients with long-standing cerebral venous hypertension may present with delayed milestones. Older children and adults usually have low flow arteriovenous fistula. These patients present with a headache and seizures.

Pathophysiology
The cardiac manifestations are due to an increase in blood flow through the cerebral arteriovenous shunt. During intrauterine life, the low resistance of the placental circulation competes with the cerebral arteriovenous shunt and, therefore, the blood flow through the shunt is not as great as that after birth. After birth, exclusion of the low resistance placental circulation results in an abrupt increase in the flow across the cerebral arteriovenous fistula.

Neurological manifestations are due to cerebral venous hypertension.

Diagnosis is done with the help of imaging modalities. Prenatal ultrasound can detect VOGMs in the third trimester of pregnancy. Contrast-enhanced CT and magnetic resonance imaging are the modality of choice for an initial assessment to locate the fistulae. Cerebral angiography is the gold standard for the evaluation of the VOGMs.

Untreated VOGMs have a very poor prognosis. Endovascular therapy is the gold standard of the treatment, and surgical treatment is associated with high mortality. Stereotactic radiotherapy has a limited role. Aggressive medical management can help postpone therapeutic embolisation until the child is above 5–6 months of age at which point of time, intervention is easier and safer. Emergency embolisation of the malformation may be necessary to reduce the shunt in patients with congestive cardiac failure that is refractory to medical therapy.

Lasjaunias et al. have described a 21 point Bicetre scale based on cardiac function, cerebral function, hepatic function, respiratory function and renal function. A score of <8 usually indicates poor prognosis and does not warrant emergency management. A Bicetre score of 8–12 is an indication for emergency management. A score of more than 12 indicates a well-preserved neonate and an attempt are made to delay the endovascular procedure by medical management.

The goal of therapy in cardiac failure patients is to arrest the congestive cardiac failure rather than to achieve complete obliteration of the shunt and then a later surgery can be performed in a staged manner to minimise the complications. Each session is limited by the volume of contrast agent delivered and how the patient tolerates the procedure. In non-cardiac failure patients, the goal is to prevent the consequences of chronic cerebral venous hypertension and promote normal cerebral development.

In our case as Bicetre score of neonate dropped from 15 to 11 emergency endovascular embolisation was planned. The anaesthetic challenges were neonatal anaesthesia, pathophysiology of the disease, remote location suite and radiation safety, haemodynamic manipulations and identifying and managing the complications.

The treatment of cardiac failure should be continued during the procedure. Both inhalational and intravenous agents can be used for induction: We used inhalation since the patient was on inotropic support. Despite potential adverse effects on intracranial compliance, ketamine is recommended for neonates in cardiac failure.
Fluid management in these cases is challenging. Neonates cannot tolerate any fluid overload because of the cardiac failure; while on the other hand, children with contracted intravascular compartment from attempts at brain dehydration may experience sudden collapse following brisk perioperative bleeding. Moreover, there should be close watch kept over the volume of dye and intra-arterial flush used during the procedure, which can put an additional load on the neonate.

During a procedure with high flow shunts, controlled decrease in blood pressure may be required to slow flow in a feeding artery before injecting glue. However, a neonate with cardiac failure may not tolerate this and is, therefore, better avoided. Invasive intra-arterial monitoring is necessary for a patient in congestive cardiac failure undergoing neuroradioembolisation to keep a close watch on the haemodynamic stability.

Fatal complications that can occur during the procedure are intracranial haemorrhage and normal perfusion pressure breakthrough due to venous hypertension. This can be largely avoided by doing a staged embolisation and maintaining normotension in the post-operative period.

Perforation of the venous sac has been reported to occur during the positioning of microcatheter and can be managed by reversal of anticoagulation and continuation of embolisation. Ischemic neurological deficits can be occasionally encountered after embolisation. Pulmonary embolisation with an embolic agent is common considering the high flow across the intracranial shunts.

Post-operatively, these patients are ventilated and sedated to maintain haemodynamic and respiratory stability. Hypertension is avoided in 1st 48 h after the procedure to prevent neurological hyperperfusion syndrome.

CONCLUSION

Management of intra-cerebral arteriovenous fistulae in the newborn is difficult. It needs to be individualised depending on the clinical parameters and the expected outcome. The Bicetre score is a useful guide to management and timely intervention. It is essential to maintain haemodynamic stability to ensure adequate cerebral perfusion pressure during the endovascular embolisation. A multidisciplinary approach involving the neonatologist, cardiologist, neuroanaesthesiologist and the interventional neuroradiologist is required to achieve a good outcome as the morbidity and mortality still remains high in the newborn period.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES