Title:
A critically ill neonate with Vein of Galen malformation requiring embolization - Importance of communication and multidisciplinary approach

Moderators: M. Saif Siddiqui M.D., Anita Akbar Ali M.D.
Institution: Arkansas Children’s Hospital, Little Rock, Arkansas.

Goals:

1. Describe the pathophysiology, clinical presentation, and diagnosis in patients with Vein of Galen Malformation (VOGM).

2. Identify perioperative concerns in patients with Vein of Galen Malformation.

3. Discuss the anesthetic management of high output cardiac failure, high intracranial pressure, and pulmonary hypertension.

4. Illustrate the importance of interdisciplinary communication in the perioperative period for these patients.

5. Design a systematic approach to address potential complications in patients with VOGM.

Case history: A full term neonate with prenatal diagnosis of Vein of Galen Malformation was transferred to a tertiary care hospital at 4:00 AM for further management. At birth, the child was hypotensive with blood pressure of 52/34 mm Hg, respiratory rate of 30/min, heart rate 65/min and pulse oximetry showing 76%, which resulted with the patient being intubated and ventilated on 100% inspired oxygen. Patient was also started on dopamine and milrinone after acquiring central venous and arterial access. Patient was transported via helicopter to a neonatal intensive care unit (NICU) of the nearby Children’s Hospital.

Questions: What is Vein of Galen Malformation? Describe the natural course of the disease? How does it present clinically? Illustrate the modalities that can be used for diagnosis? What arrangements/preparations would you do as an on call team member for perioperative optimization of the patient? What pertinent points would you consider regarding safety of the patient during transfer and handover procedure to the incoming team?

Physical Examination: On arrival to the NICU, physical examination showed hydrocephalus, pulsating anterior fontanelle, hyperdynamic precordium with pan systolic murmur, bilateral crackles, and cyanotic peripheries. Chest x ray was evident of severe cardiomegaly with bilateral pleural effusion. Arterial blood gas was 7.05/40/48/18.5
Questions: Why did the patient remain hypoxic despite being on 100% inspired oxygen? How it can be improved? What is the mechanism of developing severe cardiomegaly in these patients? What other conditions can cause a similar clinical picture?

Perioperative management: Patient was started on inhaled nitric oxide, which improved oxygenation. ABG value after 30 minutes was 7.26/53/49/18.4. MRI/MRA was performed showing non-obstructive hydrocephalus, supratentorial aneurysmal sac with multiple feeders, and widespread dilatation of venous sinuses. Retrograde flow was also noted in bilateral carotid arteries.

Questions: Discuss the significance of MRI findings. Illustrate treatment options for high intracranial pressure. Do you think the above information is adequate in order to proceed to the operating room? Is there any other information required? If yes, what difference will it make in the management and the outcome?

Perioperative management (continued): Pediatric Cardiology was consulted. An ECHO was performed revealing dilated cardiac chambers, left to right shunt through patent foramen ovale, severe pulmonary hypertension, reverse flow in coronary arteries, and arch of aorta during diastole. Coil embolization of aneurysmal feeders was scheduled in Interventional Radiology Suite. Palliative care was also involved to discuss prognosis of patient with the family.

Questions: What factors should be considered in deciding medical versus surgical treatment options? What are the perioperative concerns in a patient with Vein of Galen Malformation? What are the intraoperative anesthetic implications in a patient with pulmonary hypertension and high output cardiac failure?

Discussion:

Vein of Galen Malformation (VOGM) is one of the most complex and rare intracranial pediatric arteriovenous malformations (AVM) which poses challenging management and sometimes results in a grave outcome. Despite early and appropriate treatment, VOGM is associated with high mortality and morbidity depending upon the severity of the disease at the time of presentation.

During early embryonic life from 8th - 11th week, Median Prosencephalic vein of Markowski (MProsV) is responsible for venous drainage from cerebral tissue. It regresses later except for distal segment. This distal segment is called the Great cerebral vein or vein of Galen in adults (figure 1). Persistent and dilated MProsV forms the aneurysmal component of VOGM. The drainage of this aneurysmal malformation results into uneven distribution of circulatory volume and results in damage associated with VOGM.
In utero, placental circulation provides path of least resistance preventing damage from fluid overload in most of the cases therefore, In utero, cardiac failure is uncommon. After birth, as much as 60-70% of cardiac output is directed towards venous malformation resulting in increased venous return to cardiopulmonary systems, and ultimately leading to high output cardiac failure. The more than expected volume causes development of persistent pulmonary hypertension and persistent opening of the ductus arteriosus. These fetal circulatory shunts cause diversion of large blood volume to descending aorta and hence arterial hypoxemia and further deterioration of multi organ system. Shunt phenomena becomes worse during diastole due to retrograde flow in VOGM stealing blood away from coronary circulation resulting in cardiac ischemia and further worsening of ventricular function.

Neurological damage is another major consequence of VOGM. Large blood volume flowing through the malformation results into watershed area which increases risk of cerebral infarcts with permanent neurological deficits in severe cases.

Clinical severity depends upon the age at the time of presentation and angio-architecture of malformation. It can present at three different age groups. Neonates usually present with intractable cardiac failure and brain damage. On the other hand, infants with hydrocephalus may or may not experience cardiac failure. Older children and adults may experience headaches as the most common symptom.

Prenatal diagnosis with advanced imaging including fetal MRI and fetal Echocardiography help predict the prognosis and timely planning and referral of such critically ill patients. In the early 1960s, reported mortality rates were extremely high reaching 100% for neonates. Open neurosurgical procedure was the main stay of treatment for such complicated AVMs but introduction of endovascular techniques starting from 1980s have shown promising results.

Depending upon the time of diagnosis and initial presentation of child, appropriate treatment option can be selected. Ideally surgical intervention should be delayed till 5 months of age if condition of the patient allows. Emergent embolization is always an option in case if medical
management fails. Staged partial arterial embolization of AVM is the key. It is aimed to reverse temporary cerebral and cardiac damages gradually providing adequate time for body to adapt.

In summary, patients with Vein of Galen Malformation pose major challenge to Pediatric Anesthesiologists. Hemodynamic instability with multi-organ system failure makes them one of the most complex and critical patients to take care. Perioperative assessment and involvement of other subspecialties can help formulate an appropriate management plan to improve the quality and safety of patient care.

References:


