Case Report

Integrative Pediatrics and Child Care

A Massive Arteriovenous Malformation arising from the Aorta causing Severe Pulmonary Hypertension

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Abstract

Aortic arteriovenous malformations (AVMs) are rare vascular anomalies where the aorta communicates with adjacent venous structures, bypassing the capillary system. We report a case of a neonate born at 34 weeks gestation who presented at 3 weeks of life in respiratory distress. Echocardiography demonstrated severe pulmonary hypertension with right ventricular dilatation but an otherwise structurally normal heart. Clinical examination revealed a pulsatile abdominal mass with an audible bruit. Abdominal ultrasound was performed and it demonstrated a large cystic mass arising from the inferior vena cava (IVC) with both arterial and venous vascular flow within it. Further evaluation with magnetic resonance angiography (MRA) showed a massive arteriovenous malformation from both left and right iliac arteries, and lumbar arteries communicating with the IVC. A decision was made with interventional radiology to attempt coiling of the vascular lesion. Following this procedure his pulmonary hypertension worsened in severity, culminating in acute heart failure and multi-organ dysfunction. A subsequent interventional radiology procedure found that there was no blood flow to his bowel or abdominal viscera. Cardiac arrest followed with an unsuccessful attempt at resuscitation. This case describes a presentation of severe pulmonary hypertension initially considered to be secondary to either congenital heart disease or intrinsic pulmonary disease, but which was found to be as a result of a very rarely occurring massive aortic AVM.

Abbreviation: AVM: Arteriovenous Malformation; AVF: Arteriovenous Fistula; IVC: Inferior Vena Cava; CPAP: Continuous Positive Airway Pressure; NIV: Non Invasive Ventilation; MRA: Magnetic Resonance Angiography; PVR: Pulmonary Vascular Resistance; PBF: Pulmonary Blood Flow; SVT: Supra Ventricular Tachycardia

Introduction

Arteriovenous malformations (AVMs) are micro-fistulas consisting of multiple arterial feeders which join via a nidus to draining veins. Arteriovenous fistulas (AVFs) are macro-fistula, direct shunts between large arterial and venous channels without an intervening capillary bed [1]. These vascular malformations occur due to dysregulation in morphogenetic vascular development [2]. The most common site for neonatal vascular malformations is the liver, followed by the brain and lung [3]. Diagnosis of AVMs can be difficult in the neonatal period as their

clinical presentation is very variable [4]. As AVMs are extremely rare, they are far down the list of potential causes of severe pulmonary hypertension in the neonatal period. Common known causes of severe pulmonary hypertension in the neonatal period direct the physician towards focusing on identifying an underlying cardiac or respiratory aetiology. This case highlights an extremely rare cause of pulmonary hypertension.

Case Report

A male neonate born at 34 weeks gestation was

transferred to the pediatric intensive care unit (PICU) at our institution at 22 days of life in severe respiratory distress. His early clinical course in the neonatal intensive care unit (NICU) was marked by intermittent bouts of tachypnea requiring nasal continuous positive airway pressure (CPAP), and intermittent supraventricular tachyarrhythmias (SVTs) responding to adenosine 0.2 mg/kg as needed. At 15 days old he underwent an echocardiogram to investigate for structural heart disease. This showed mild mitral regurgitation and mild branch pulmonary stenosis but otherwise structurally normal heart, with good ventricular function. A week later, the neonate had a clinical deterioration with increased respiratory distress and an episode of pyrexia, and was transferred for further evaluation and management. On arrival to the PICU the infant was in severe respiratory distress, with a respiratory rate of 120 breaths per minute, oxygen saturations of 95% on 1 liter oxygen via nasal prongs, and significant sub-costal recession. He had a heart rate of 180 beats/minute in normal sinus rhythm, blood pressure of 52/24 mmHg, capillary refill time less than 2 seconds, and lactate 2.2 mmol/l. Physical examination revealed a palpable abdominal mass, arising from the pelvis, with an audible bruit. There were no other abnormal features. Blood tests showed elevated liver enzymes but otherwise were normal. Chest X-ray showed cardiomegaly, pulmonary oedema, and small bilateral pleural effusions. Repeat echocardiography showed tricuspid regurgitation with a gradient of 65mmHg, an engorged main pulmonary artery, moderate right ventricular and right atrial dilatation, with preserved left ventricular function, without anatomic abnormality. Noninvasive ventilation (CPAP) was commenced along with intravenous furosemide (1mg/kg twice daily) and oral sildenafil (1mg/kg three times daily) to offload the pulmonary circulation.

Over the next 24 hours the patient clinically improved with reduced work of breathing. An abdominal ultra sonography exam in light of the physical findings and a computed tomography of the thorax to evaluate for intrinsic pulmonary disease were planned. Abdominal ultra sonography showed a large cystic mass arising from the inferior vena cava (IVC) with arterial and venous flow within, suggestive of an AVM, and the decision was made to proceed with a magnetic resonance angiogram (MRA) of the abdomen, chest and pelvis to further evaluate the mass. MRA (Figure 1 and Figure 2) revealed a complex AVM measuring 5.1cm in diameter at its widest, between the distal IVC and the aorta at its bifurcation with the common iliac arteries, with feeding vessels noted from

the lumbar and internal iliac arteries respectively. There was normal blood flow to the lower limbs.

A multidisciplinary meeting was organized to discuss potential surgical and interventional radiology management options and it was felt that radiological coiling of the AVM was the most suitable option. As this service is not available in the Republic of Ireland, transfer was arranged to a quaternary paediatric hospital in the United Kingdom for attempted radiological coiling. In the interim, the neonate's respiratory status deteriorated, necessitating intubation and mechanical ventilation. He was commenced on a milrinone infusion (0.5 micrograms/kg/min), spironolactone (1mg/kg twice daily), and sildenafil was discontinued. The patient was transferred to the quaternary paediatric centre in the United Kingdom after 6 days in our ICU. Following transfer, the patient underwent an attempted radiological coiling with almost 50 individual coils, which appeared successful in reducing flow into the AVM. Over the ensuing 72 hours, there was significant cardio respiratory compromise with supra-systemic pulmonary hypertension, and acute cardiovascular failure requiring maximal inotropic support. This clinical deterioration necessitated a repeat interventional radiological procedure to determine if the shunt was still open. This procedure confirmed that the shunt was closed, but it also demonstrated that there was no blood flow to the bowel or abdominal viscera. He had a cardiac arrest shortly after and resuscitation was unsuccessful.

Discussion

Arteriovenous malformations are abnormal connections between arteries and veins without intermediary capillary beds. These lesions can occur anywhere in the body and clinical symptoms and signs are related to both the location and dimensions of the AVM [5]. Cases of massive aortic AVMs giving rise to suprasystemic pulmonary hypertension are sparsely reported in the literature [4]. Most documented cases of symptomatic AVMs refer to those occurring within the cerebral or hepatic circulations. AVMs result in the shunting of blood directly from the high pressure arterial system to the low pressure venous system. In large aortocaval vascular lesions such as the one described large volume blood steal directly from the arterial system into the venous system resulting in pulmonary circulatory overload, pulmonary hypertension, and high output cardiac failure. Severe suprasystemic pulmonary hypertension with concomitant right ventricular dysfunction is a life threatening condition which must

Figure 1: Magnetic Resonance Angiographic Image of Pelvic Arteriovenous Malformation

Figure 2: Magnetic Resonance Angiographic Coronal T2 Image of Arteriovenous Malformation

be managed expertly and urgently all while conducting investigations to determine the underlying cause [6]. Pulmonary hypertension in neonates is much more commonly ascribed to lung hypoplasia, pulmonary parenchymal pathology, congenital cardiac disease, and sepsis. The key feature of pulmonary hypertension in these disease states is an elevated pulmonary vascular resistance (PVR). There are few instances where increased pulmonary blood flow (PBF) result in neonatal hypertension, one of which is the presence of a massive AVM [7]. There have been multiple case reports in the literature of neonatal pulmonary hypertension secondary to cerebral, hepatic, and pulmonary AVMs, but only one of neonatal suprasystemic pulmonary hypertension due to a massive retroperitoneal aortocaval AVM [4]. The supportive management of pulmonary hypertension in this case included NIV CPAP, furosemide (1mg/kg twice daily), and spironolactone (1mg/kg twice daily) to aid in reducing circulatory overload. While inhaled nitric oxide therapy is used extensively in treating neonatal pulmonary hypertension secondary to raised PVR, caution is exercised with pulmonary hypertension due to increased PBF as there is a theoretical risk of worsening circulatory overload [7]. Ultimately to treat pulmonary hypertension from a massive AVM the lesion must be occluded to reduce pulmonary blood flow. The options for therapeutic management of a massive aortocaval AVM are surgical resection and radiological coiling [8].

Given the potential for massive intra-operative blood loss with open surgery it was felt that the risk-benefit favoured radiological coiling of the AVM. After what was believed to initially be a successful closing of the massive shunt, the patient deteriorated clinically, requiring intensive respiratory and cardiovascular support. Repeat angiography showed no blood flow to bowel or abdominal viscera. The cause of mesenteric ischaemia was an embolic shower from the coil mass. Thromboembolic complications from endovascular coiling are relatively uncommon with an incidence of 2 – 11% [9].

In conclusion we present the case of a massive aortocaval AVM producing supra-systemic pulmonary hypertension, ultimately progressing to acute heart failure. Our case demonstrates that when an obvious cause for severe neonatal pulmonary hypertension cannot be identified investigations should rapidly move towards outruling congenital vascular malformations as management of the underlying cause should coincide with critical care support. This case highlights the importance of a comprehensive physical examination as detection of the abnormal pulsatile mass on abdominal palpation altered our differential diagnosis, which was subsequently confirmed on abdominal ultra sonography. If ultra sonography identifies the presence of a large AVM, MRA should take place to further delineate the anatomy of the lesion and to plan appropriate therapeutic

intervention.

Contributors Statements

Patrick John Kennelly: Dr Kennelly conceptualized the case report, and drafted the manuscript, both initial and final versions.

David Rea: Dr Rea analysed the radiological images for this case report, and developed the final presentation of these images with appropriate anatomical markings.

Suzanne Crowe: Dr Crowe conceptualized and supervised the writing of the article, and critically reviewed the manuscript prior to submission.

All Authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

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