Case report

Right ventricular inflow obstruction from massive fungal vegetation presenting as neonatal circulatory collapse

Luca A. Vricella, Sachin Khambadkone, Robert Yates, Victor T. Tsang*

Cardiothoracic Unit, Great Ormond Street Hospital for Children NHS Trust, Great Ormond Street, London WC1N 3JH, UK

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Abstract

Occurrence of neonatal circulatory collapse imposes effective differential diagnosis and expeditious therapeutic intervention. We report a case of neonatal cardiogenic shock, caused by a massive intra-cardiac fungal vegetation.

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1. Case report

A 3.5 kg, term-neonate female was admitted to the neonatal intensive care unit shortly after vaginal delivery with low Apgar score and birth asphyxia, resulting in protracted seizure activity. The newborn was mechanically ventilated for 48 h, and eventually discharged on barbiturate therapy, with a diagnosis of mild hypoxic encephalopathy. On the 20th day of life, she was brought to her paediatrician with increasing lethargy and emesis. She was transferred to Great Ormond Street Hospital where she presented in extremis, with evidence of poor tissue perfusion, deep cyanosis (oxymetric saturation of 35%) and faint pulses. Resuscitation was begun immediately, with endotracheal intubation and administration of intravenous fluids. Dopamine infusion, as well as several boluses of adrenaline and noradrenaline, were followed by an unsustained clinical response. Broad-spectrum parenteral antibiotics were started concomitantly.

Initial laboratory evaluation was significant for profound metabolic acidosis (pH = 6.8, serum lactate = 22 mmol/l) as well as leucocytosis (white blood cell count of 55 × 10^3/ml) and thrombocytopenia (platelet count 38 × 10^3/ml). A transthoracic echocardiogram (Fig. 1) revealed an extensive, lobulated mass extending across the tricuspid valve, as well as a small patent ductus arteriosus and a patent foramen ovale (PFO) with right-to-left shunting.

The newborn was brought emergently to the operating room, where the large mass was excised through a right atriotomy (Fig. 2). Cardiopulmonary bypass with bi-caval cannulation, induced fibrillation (26 min) and mild normothermia (core temperature of 34.0°C) were utilised. The 6 cm tumour was attached to the inlet portion of the inferior vena cava, as well as both Eustachian and tricuspid valves. Pathological evaluation of the specimen disclosed fibrin, inflammatory cells and fungal elements consistent with Candida albicans.

The postoperative course was marked by refractory pulmonary hypertension requiring nitric oxide therapy as well as prolonged mechanical ventilation. A ventilation-perfusion scan was unremarkable, while persistent right-to-left shunting through the PFO and mild-to-moderate tricuspid regurgitation were noted on follow-up echocardiography. Magnetic resonance imaging of the brain revealed multiple small, scattered embolic foci. The patient was discharged on the 29th postoperative day on long-term intravenous anti-fungal therapy (flucytosine and amphotericin B), and was doing well at latest follow-up, 4 months post-discharge.

2. Comment

Circulatory collapse and cyanosis in neonates demands
immediate differentiation between cardiac and non-cardiac causes of shock. Though the latter (sepsis in particular) occurs more frequently, cardiogenic causes should be ruled out immediately. Two-dimensional echocardiography will reliably diagnose a variety of malformations with ductus-dependent circulation, as well as other more rare causes of cardiogenic shock, such as intra-pericardial tumours or coronary anomalies.

Atrio-ventricular valve obstruction from tumours is certainly a rare cause of collapse in the newborn, though not an exceedingly rare cause of in utero fetal demise [1]. Typical presentation of a cardiac tumour is that of right or left ventricular failure, atrioventricular heart block or embolic phenomena. Though operative intervention results in acceptable operative outcomes and long-term palliation or cure [2], some authors have advocated non-operative management of non-haemodynamically compromised patients, having observed with encouraging frequency spontaneous tumour mass regression over time [3–5].

Rare reports have described intra-cardiac masses in the setting of fungal septicemia and embolic phenomena, with successful non-operative treatment [6]. The initial event causing development of this massive vegetation can be attributed to an indwelling umbilical vein catheter with positive cultures for *C. albicans* utilised during this neonate’s initial intensive care unit admission. This particular presentation mandated immediate intervention because of haemodynamic compromise; aggressive surgical management in the setting of clinically evident septic embolisation may be justified as well. Persistent severe pulmonary hypertension in the postoperative period was in our opinion most likely the result of pulmonary microembolisation; maintaining a PFO was most certainly beneficial in the perioperative management of this newborn.

Therapeutic advances and more aggressive neonatal intensive care have resulted in improved survival, as well as an increasing population of low-birth weight premature neonates who often require prolonged use of indwelling venous catheters. Meticulous surveillance and early pharmacological intervention are imperative in order to avoid severe complications from virulent microorganisms [7].

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**References**