Case report - Congenital

Methemoglobinemia masquerading as pulmonary hypertension

Raj Gopal Menon*, Shobha Menon, John Valliyathu, Taha Al Delamie

Department of Cardiothoracic Surgery, The Royal Hospital, P.O. Box 1331, PC 111, Seeb, Oman

Received 4 July 2003; received in revised form 7 September 2003; accepted 10 September 2003

Abstract

Pulmonary arterial hypertension is known to occur postoperatively after closure of VSD. It results in a low cardiac output state with need for ventilation and frequent bagging. Methemoglobinemia, a condition associated with toxic ingestions, has been described in this setting. Methemoglobinemia is known to cause cyanosis and hypotension. We report a critically ill infant with significant methemoglobinemia after VSD closure.

Keywords: Methemoglobin; Cardiac surgery; Pulmonary hypertension

1. Introduction

Pulmonary hypertensive crisis (PHC) is known to occur in infants after closure of VSD. It results in a profound fall in cardiac output (CO) with fall in arterial oxygen saturations (SaO₂). At times, this situation may be irreversible in spite of urgent intensive management. Methemoglobin (MHb) has minimum oxygen carrying capacity. Hence methemoglobinemia results in cyanosis, systemic acidosis and severe hypotension. Thus, methemoglobinemia presenting after cardiac surgery can be mistaken for pulmonary hypertension. Though life threatening, methemoglobinemia can be reverted quickly. The knowledge of such a presentation can speed up recovery.

2. Case report

AR, a 1-month-old boy was diagnosed to have multiple VSDs, large ASD, coarctation of aorta, severe pulmonary artery hypertension (PAH), and G6PD deficiency. He presented with severely compromised cardiovascular function, needing ventilation and multiple ionotropes. Since it was not possible to do a complete correction in such a critical situation, pulmonary artery (PA) banding and repair for coarctation of aorta was done on 23 July 2002, to tide over the crisis. His postoperative course had been uneventful. During follow-up, the pulmonary artery pressure (PAP) continued to rise (95 mmHg), the baby needed three hospital admissions for congestive cardiac failure and respiratory tract infections. Cardiac catheterization on 29 October 2002 showed a gradient of 10 mmHg across the repaired coarctation site, equal LV and RV pressures (120 mmHg) and PAP 80 mmHg. On 10 November 2002, he was operated for removal of PA band, autologous pericardial patch reconstruction of PA, closure of four VSDs (one subaortic, three muscular) and large secundum ASD. After he was weaned off cardiopulmonary bypass (CPB) unevenly, the PAP was 90 mmHg (same as systemic). Hence, glicerytrinatrate (GTN) infusion was started to reduce the PAP. Postoperatively, he was electively ventilated; dopamine (5 μg/kg/mt) and adrenaline (0.03 μg/kg/mt) infusions were started. The next day, as the hemodynamics, SaO₂ and arterial blood gases (ABG) were optimum; it was decided to slowly wean him off the ventilator. However, when the oxygen flow (FiO₂) was reduced from 80 to 75%, SaO₂ dropped to 90%, ABG showed partial pressure of oxygen (PO₂) of 70 and the systemic arterial pressure (SAP) fell to 58/36. After increasing tidal volume (10 ml/kg) and FiO₂ (100%), PO₂ improved to 142 and SaO₂ to 96%. The SAP improved to 64/40 mmHg after doubling the dose of ionotropes. However, with the slightest reduction of FiO₂ (95%), SaO₂ dropped to 64% and SAP fell to 50/32 mmHg, even when the PO₂ was within normal limits. The only way...
we could maintain the SaO\textsubscript{2} 80\% and SAP 60 mmHg systolic was by frequent bagging. The baby had developed a tinge of cyanosis with peripheral temperature 34 °C. Since it was an acute low CO state in an infant after VSD, ASD closure, the patient was managed as a case of post-operative PHC. The patient was kept warm (wool blankets, over head warmer), sedated and paralyzed using Rocuronium, Fentanyl and Dormicum infusions. The CVP was kept at 12 to increase preload. Dobutamine and calcium infusions were added to improve SAP. Despite 100% FiO\textsubscript{2} and higher doses of inotropes, SaO\textsubscript{2} and SAP kept falling. This low CO state persisted even after repeated correction of metabolic acidosis and additional inotropes. All instrument variables (monitor, pulse oximeter, ABG analysis, ventilator) were rechecked and instruments changed, however, the saturations continued to drop.

Finally, it was the persistence of high PaO\textsubscript{2} levels (242) with low SaO\textsubscript{2} (70–90\%), that suggested the diagnosis of methemoglobinemia. The methemoglobin level was checked and was found to be 10.6 g/dl.

GTN infusion was stopped immediately. A bolus of 1 ml/kg methylene blue was given, after which the SaO\textsubscript{2} and SAP improved dramatically. It was then possible to come down on the FiO\textsubscript{2} and inotropes. In the next 24 h, it was possible to slowly reduce the ventilatory support. The patient was only on dopamine (3 \textmu g/kg/min), and the overall condition of the patient improved. Pneumonitis further delayed weaning, though a week later he was extubated and remained well.

3. Discussion

Methemoglobin (MHb) may arise from genetic [1], dietary [2], idiopathic causes [3] and toxic agents [4]. Underlying lung, heart and blood disorders can exacerbate the toxicity of MHb [3]. Symptoms vary from headache, cyanosis, hypotension, to coma/death and may not correlate to the MHb concentrations.

Postoperative PHC is described as a syndrome of hyperacute increase in the PAP, followed by profound reduction of CO and fall in SaO\textsubscript{2}; in infants ventilated after operation for congenital cardiac anomalies with PAH. Acute hypoxia and catecholamine infusions are known post-operative risk factors for PHC [7].

Since this infant had severe PAH even after the second surgery, the persistent low SaO\textsubscript{2}, and low CO were initially attributed to PHC; and treated accordingly. The patient did not have methemoglobinemia after his first surgery even though he was G6PD deficient. Hence methemoglobinemia was not suspected to be the reason for low CO after the second operation.

Metabolic acidosis [3], anemia [3] and CPB [5] are known to further accentuate the clinical effects of MHb. The presence of all three factors in this infant made his condition extremely critical. This presentation of methemoglobinemia is unique and not mentioned before in literature.

The foremost step to reduce MHb levels is to remove the oxidizing agent. In this case, the glyceryltrinitrate infusion was the culprit. Infusion of methylene blue, cytochrome P-450 inhibitor: cimetidine and exchange transfusions are the conventional methods to correct methemoglobinemia [3]. The use of methylene blue in neonates can cause Heinz body hemolytic anemia [6]. In this baby, methylene blue not only reversed the clinical situation dramatically (MHb < 2 g/dl within 48 h); also serology showed no Heinz body hemolytic anemia.

Cyanosis and low CO with normal PO\textsubscript{2} levels should alert the clinician to the presence of methemoglobinemia. All modern ABG analyzers do give a reading of methemoglobinemia levels. One has only to be alert about this situation.

References