Case Report

Undiagnosed intraoperative methaemoglobinaemia

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ABSTRACT

Methaemoglobinaemia is a rare but potentially dangerous haemoglobinopathy that is often underdiagnosed. It is one of the causes for unexplained cyanosis with dark-coloured blood, especially in the absence of cardiac or pulmonary pathology. Not uncommonly so, it is an incidental perioperative finding in cases of dark-coloured blood not improving with oxygen in apparently acyanotic patients. The present case report is of a child with deaf-mutism posted for cochlear implant surgery who presented with 'chocolate-coloured blood' in the surgical field, despite blood gas analysis showing a normal partial pressure of oxygen.

Key words: Dark-coloured arterial blood, haemoglobinopathy, methaemoglobinaemia

INTRODUCTION

Anaesthesiologists in general tend to concentrate on the cardiorespiratory system for the differential diagnosis of cyanosis, especially in paediatric patients. Haemoglobinopathies, especially methaemoglobinaemia, are rare but important cause of dark-coloured blood with or without clinically apparent cyanosis.[1] The incidence is rare and not known. The prevalence is difficult to determine because of a wide array of presentations ranging from asymptomatic underdiagnosed cases to potentially fatal cases.[1]

CASE REPORT

A 7-year-old child weighing 23 kg presented to the ENT Department with deaf-mutism since birth. Developmental milestones and intelligent quotient were normal. Cardiorespiratory system and airway examination were normal. Laboratory reports were within the normal limits. He was scheduled to undergo cochlear implant surgery.

On the day of surgery, preoperative heart rate was 100 beats/min, SpO_2 93% on room air and blood pressure 100/60 mmHg. He was premedicated with intravenous (IV) glycopyrrolate (5 μg/kg), IV fentanyl (2 μg/kg) and IV ondansetron (0.1 mg/kg). Anaesthesia was induced with IV propofol 2 mg/kg and IV atracurium 0.5 mg/kg, followed by intubation with a 6-mm ID cuffed endotracheal tube. Anaesthesia was maintained with oxygen, air and sevoflurane (1 minimum alveolar concentration).

Following incision, the surgeon noticed 'dark-coloured blood' in the surgical field, and a decrease in oxygen saturation from 94% to 89% was noticed by the anaesthetist. Immediately, the FiO_2 was increased to 100% and the position of the endotracheal tube was checked. Good bilateral air entry and a good capnographic waveform confirmed optimum ventilation. An immediate arterial blood gas (ABG) sample was drawn that appeared 'dark coloured', but had the following results: PaO_2: 324 mmHg, PCO_2: 22 mmHg, HCO_3: 24 mmol/L and pH: 7.43.

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Undiagnosed haemoglobinopathy was strongly suspected and surgery was postponed pending further evaluation. Co-oximeter analysis revealed 21% methaemoglobin. Haemoglobin (Hb) electrophoresis and G6PD levels were normal. Erythrocyte nicotinamide adenine dinucleotide (NADH)-dependent methaemoglobin reductase level was low at 20.5 IU/g Hb (normal 35 ± 5 IU/gm Hb). A history of consanguinity was revealed on further questioning. He was promptly started on oral methylene blue (100 mg thrice daily) and oral ascorbic acid (200 mg/day) for 1 week.

Surgery was rescheduled following 1 week of methylene blue therapy, after which, his methaemoglobin levels decreased to 1.2% and pulse oximetry showed 100% saturation.

Pre-operative vitals were normal (heart rate: 100 beats/min, oxygen saturation: 100% and blood pressure: 96/65 mmHg). He was premedicated with IV fentanyl and anaesthesia was induced with IV propofol. Endotracheal intubation was facilitated with IV atracurium, while 100% oxygen and sevoflurane were used for maintenance. Methylene blue was kept ready in the operating theatre. Surgery was uneventful and he was extubated after regaining full consciousness. Post-operative oxygen saturation was 100% and ABG was normal.

DISCUSSION

Methaemoglobinaemia is one of the rare haemoglobinopathies that may present with 'dark-coloured arterial blood' with or without clinical signs and symptoms. The pathology implicated is oxidation of ferrous ion ($Fe^{2}+$) of the Hb molecule to the ferric state ($Fe^{3}+$) that hampers oxygen-binding capacity of Hb molecule and hence oxygen delivery to tissues.[2] Normally, the amount of methaemoglobin is <1%.[3,4] Clinical signs and symptoms are produced when this percentage increases. Levels up to 20% may produce slight discolouration or cyanosis. Levels of 25%–50% can produce headache, dizziness or syncope. Levels of 50%–70% can result in arrhythmias, delirium, seizures and coma. Levels >70% result in circulatory collapse and acidosis and may be fatal.[5]

Both congenital and acquired aetiologies are reported. The congenital variety usually occurs with NADH methaemoglobin reductase deficiency and shows autosomal recessive inheritance.[6-8] The acquired variety occurs secondary to exposure to certain medications such as acetaminophen,[9] prilocaine and EMLA®,[10] ibuprofen, metoclopramide, amyl nitrite, silver nitrate, nitroglycerin, nitric oxide[11,12] and dapsone.[3]

In the present report, the child probably had congenital variety of the disease as evidenced by low levels of NADH methaemoglobin reductase enzyme and a family history of consanguinity with a normal elder sibling. Diagnosis is based on co-oximetric analysis of methaemoglobin percentage.[7] Reduced levels of NADH methaemoglobin reductase enzyme are seen in the congenital variety. ABG is generally normal. The condition has to be distinguished from more common causes of cyanosis such as cardiorespiratory disease or sulfhaemoglobinemia (that is unresponsive to methylene blue).

Treatment consists of methylene blue (1–2 mg/kg IV) as 1% solution given over 5–10 min[13] or oral 60 mg 3–4 times a day. The rationale is that activation of NADH methaemoglobin reductase restores the reduced form of iron in Hb molecule. Levels of methaemoglobin are reported to fall as early as 30–60 min after administration of methylene blue.[13] Supportive measures may include ascorbic acid, hyperbaric oxygen therapy and in unresponsive cases exchange transfusion.[14] Genetic counselling is warranted in families with a history of methaemoglobinaemia or other haemoglobinopathies.

Undiagnosed cyanosis or its indirect evidence (e.g., dark-coloured blood) can cause alarm and unwanted panic in the operating theatre. Oxygenation and ventilation are the usual suspects and a misplaced endotracheal tube is the most blamed factor. But, in the presence of optimal ventilation with 100% oxygen, dark-coloured blood and low oxygen saturation, should raise suspicion of rare entities such as methaemoglobinaemia. Vigilance and a high index of suspicion is warranted on the part of the anaesthetist while dealing with pre-anaesthetic examination or perioperative management of children with not so apparent cyanosis in the face of a normal cardiorespiratory profile.

Declaration of patient consent

The authors certify that the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal patient identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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