A Case Report of Central Cyanosis
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Abstract
Congenital methemoglobinemia is a rare condition of central cyanosis. We report a case 14 year old boy with normal cardiovascular and respiratory function, with peripheral cyanosis. He was diagnosed to have methemoglobinemia based on the findings of polycythaemia. This should be rare cause of central cyanosis which needs to be ruled out.

Keywords: Methemoglobinemia; Congenital; Cyanosis

Introduction

Methaemoglobinaemia is an uncommon etiology of cyanosis, but one that demands prompt diagnosis and treatment [1,2]. History, physical examination, bedside diagnostic techniques and laboratory confirmation are all important in the evaluation. However, in the absence of significant history, mild cyanosis can be easily missed in dark skinned individuals during the pre-anesthetic checkup. We report a case of methaemoglobinaemia which was first diagnosed in the intraoperative period due to appearance of chocolate brown colored blood after surgical incision.

Case Report

We report a case 14 year old boy with history of absence of right testicle since birth, and diagnosed to have undescended right testis. No other symptoms related to cardiovascular and respiratory system. On examination he had bluish discoloration of lips and tongue and peripheral cyanosis. Growth normal and he was moderately built, not aware of peripheral cyanosis. Other systems in particular cardiovascular and respiratory were essentially unremarkable, no organomegaly. Right testis not palpable in the inguinal canal and scrotum. Patient planned for right orchidopexy under spinal anesthesia, in the operation theatre testicle not found in the inguinal canal and scrotum. Patient planned for right orchidopexy under spinal anesthesia, in the operation theatre his Pulse 85 beats/min, NIBP -120/70 mmhg and ECG was normal.

When the incision was made in right groin dark chocolate colored blood oozed, on checking pulse oximetry SpO2 was 89%. Patient was dark complexion hence preoperatively cyanosis could not be made out. But on close observation bluish discoloration of tongue and nail beds noted. Even after administering 100% O2 the saturation did not raise above 93% and no improvement in the color of blood made out. An arterial blood gas analysis revealed normal report. Since the patient was hemodynamically stable, the surgery was completed without complications. Patient shifted to recovery room, where a detailed history was taken regarding the cardiorespiratory, recurrent respiratory infections and previous hospitalization and exposure any chemicals or family history of similar condition. Since patient was not aware of peripheral cyanosis. Other systems in particular cardiovascular and respiratory function, with peripheral cyanosis. We report a case of methaemoglobinaemia which was first diagnosed in the intraoperative period due to appearance of chocolate brown colored blood after surgical incision.

Laboratory investigations revealed Hemoglobin-14.6 gms% (normal for his age is 11-14), RBC–5.40X106 Cells/mm3, PCV–43.8%, MCV, MCHC, MCH are normal. Platelet and differential count within normal limits. RBC morphology normocytic and normochromic. ECG and ECHO are normal, based on the above investigations and examination methemoglobinemia was suspected and hemoglobin electrophoresis is planned. USG revealed iso echoiec lesion just below the deep inguinal ring, no other abnormalities noted. Patient awareness of peripheral cyanosis. Other systems in particular cardiovascular and respiratory were essentially unremarkable, no organomegaly. Right testis not palpable in the inguinal canal and scrotum. Patient planned for right orchidopexy under spinal anesthesia, in the operation theatre his Pulse 85 beats/min, NIBP -120/70 mmhg and ECG was normal.

Hemoglobin electrophoresis revealed adult hemoglobin of 95% and no M band hemoglobin (Figure 1).

Discussion

Methaemoglobinaemia is a condition in which the iron within hemoglobin is oxidized from the ferrous (Fe2+) state to the ferric (Fe3+) state, resulting in the inability to transport oxygen and carbon dioxide [3-5]. Methaemoglobinaemia occurs when methemoglobin levels is more than 2%. The failure of 100% oxygen to correct cyanosis is suggestive of methaemoglobinaemia [6,7]. Diagnosis is based upon central cyanosis unresponsive to oxygen therapy decreased measured oxygen saturation in presence of a normal PaO2. Since methemoglobin has absorption characteristic similar to that of deoxyhaemoglobin, its presence in blood lowers the saturation as read on the pulse oximeter. The saturation reported on the arterial blood gas is based on the partial pressure of dissolved oxygen and assumes no abnormal hemoglobin is present therefore the reported oxygen saturation in arterial blood gas analysis is higher than that measured with the pulse oximeter [8].

Methaemoglobinaemia can be acquired secondary to exposure to oxidant agents; few such agents commonly used in anesthesia practice include nitroglycerine, local anesthetics like lidocaine and prilocaine [9,10]. Congenital causes of methaemoglobinaemia include deficiency

![Cyanotic tongue.](image)

**Figure 1:** Cyanotic tongue.

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of enzyme NADH cytochrome-b5 reductase (autosomal recessive),
cytochrome-b5 deficiency (autosomal recessive) or hemoglobin M
disease due to globin chain mutation (autosomal dominant) [11,12].

Conclusion

To conclude our case is a rare case of central cyanosis without
having cardiorespiratory symptoms and exercise intolerance, need
to be vigilant in pre-operative evaluation of cases and importance of
general physical examination to be kept in mind.

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