RESEARCH PAPER | Medical Science | Volume : 4 | Issue : 1 | Jan 2014 | ISSN - 2249-555X

A case of methemoglobinemia

KEYWORDS: Acquired and hereditary methemoglobinemia, methylene blue

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ABSTRACT *Methemoglobinemia is a rare cause of cyanosis characterised by increased concentration of methemoglobin in the blood with an altered state of haemoglobin in which the ferrous form of iron is oxidized to ferric state rendering heme moiety incapable of carrying oxygen. We are presenting a young boy admitted with giddiness and cyanosis , in whom cardiopulmonary causes of cyanosis were excluded. Clinical cyanosis and low measured oxygen saturation in presence of normal arterial oxygen tension was highly suggestive of Methemoglobinemia. Methemoglobin level was more than 20 %. He was successfully treated with nasal oxygen and ascorbic acid.*

Introduction –
If not treated adequately, methemoglobinemia may be lethal and therefore should be considered in all cyanotic patients, without any prior history of cardiac or respiratory problems, who remain unresponsive to oxygen therapy.

Case Report-
18 years old boy, came to emergency department with acute onset giddiness and dyspnoea over a period of one hour. There was no history of any medical illness in past as well as in family. He denied consumption of any medication or any illicit drug.

On examination, he was conscious , oriented but drowsy. His pulse rate was 100/min, respiratory rate 32/min and blood pressure was 138/80 mm of Hg. He had marked peripheral as well as central cyanosis. Systemic examination revealed no abnormality. His blood oxygen saturation measured by pulse oxymetry was 65% . Blood drawn for laboratory testing was chocolate brown . Arterial PH was 7.44, PCO2- 28.5mm Hg and PO2 was 138 mm Hg on nasal oxygen 2 L/min. Haemoglobin was 13.9 gm %. Rest of the metabolic laboratory parameters were within normal limits. He was HIV negative. Electrocardiogram showed sinus tachycardia. Chest radiogram and echocardiography findings did not show any abnormality. G6PD level was normal.

Respiratory and cardiac causes of cyanosis were excluded. In the presence of clinical cyanosis and low measured oxygen saturation with normal arterial oxygen tension , diagnosis of methemoglobinemia was suspected clinically. So his blood was tested for methemoglobin level which was more than 20 %. (normal 0- 1.5 % of total Hb). He was started with high flow oxygen by mask and oral ascorbic acid 500 mg daily. By next morning, patient's symptoms resolved completely. His cyanosis decreased over a period of two days. On third day of admission, repeat methemoglobin level was 16 %. He was discharged with full recovery. This may indicate the possibility of hereditary methemoglobinemia.

Discussion –
Methemoglobinemia is characterised by increased concentration of methemoglobin in the blood with an altered state of haemoglobin in which the ferrous form of iron is oxidized to ferric form. Methemoglobin is not capable of carrying oxygen and thus causes varying degree of cyanosis.

This condition may be genetic or acquired. Hereditary form is a rare recessively inherited disorder due to deficiency of an enzyme nicotinamide adenine dinucleotide (NADH) cytochrome b_5 reductase. Type I deficiency is limited only to erythrocytes and cyanosis is the only symptom while type II form is more severe with deficiency of enzyme in all tissues and with strong neurological component.(1)

Acquired methemoglobinemia is more common than the genetic form and caused by many chemical agents and drugs. In a retrospective study of 138 cases of acquired methemoglobinemia, patients' ages ranged from 4 days to 86 years and dapsone was the most common etiology. 20% benzocaine spray was the causative agent in 5 cases with severely elevated levels of methemoglobin . 11 paediatric patients had methemoglobin either due to exogenous exposure like drugs or because of serious illness like gastrointestinal infection and dehydration. 94% patients were anaemic.(2)

Nitrites are also common offenders. Outbreaks of methemoglobinemia have been reported due to nitrite poisoning from water contamination. (3)

The optimum treatment of methemoglobinemia is stopping the offending drug , adequate oxygen delivery and appropriate antidotal therapy. For methemoglobinemia due to drug exposure, methylene blue infusion is the first line therapy. It is important to rule out G6PD deficiency in these patients because if there is a co existent G6PD deficiency , methylene blue is ineffective and its use may result in hemolysis. Effectiveness of N –acetyl cysteine and cimetidine are also studied as a treatment options for methemoglobinemia. (4) (5). Exchange transfusion is reserved for the patients in whom methylene blue therapy is ineffective. Automated red blood cell exchange may be superior to manual exchange transfusion for treatment of patients with high-risk acquired methemoglobinemia. (6)

Most of the type I hereditary methemoglobinemia have no clinical consequences and need not be treated or treatment can be directed at improving the poor cosmetic effect of persistent cyanosis with minimum amount of drugs to give satisfactory clinical results (7) , In such patients oral methylene blue or ascorbic acid 200 -500 mg daily can reverse cyanosis.. One study done in animals , concluded that vitamin C as well as vitamin E can reduce oxidative induced methemoglobin formation in vitro in dose dependent manner, separately or in combination and can be used as a alternative medication. (8)

In this case report, cause of methemoglobinemia was not established .Patient responded well to oral ascorbic acid and high flow oxygen therapy and became asymptomatic in few hours.

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Conclusion –

Severe methemoglobinemia is a medical emergency and high level of suspicion is required to make a diagnosis. It should be considered in all patients with cyanosis that is refractory to oxygen therapy , without any prior history of cardiac or respiratory problems.

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