Case Report

Peri operative Management of a Patient with Methaemoglobinaemia Posted for Caesarean Section

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Abstract

Methaemoglobinaemia is an uncommon but potentially serious disorder. Stress of pregnancy in a case of congenital methaemoglobinaemia leads to increased Methaemoglobin levels. This at best leads only to aggravation of maternal symptoms like dyspnoea and at worst results in obstetric complications compromising maternal and foetal safety. We present a case of recently diagnosed congenital Methaemoglobinaemia who underwent caesarean section with successful outcome.

Key words: Methaemoglobinaemia, Peri operative Management, Pregnancy

Introduction

Methaemoglobinaemia is an uncommon but potentially serious disorder which results in impaired oxygen delivery. Oxidative stress in these patients leads to excess production of free radicals and oxidative damage to cellular membranes and DNA [1]. This may result in uteroplacental insufficiency leading to IUGR, threatened abortion, abruptio placentae, PIH, preterm labour and foetal distress [1, 2]. The literature relating to congenital Methaemoglobinaemia in pregnancy is sparse [3]. We present a case of recently diagnosed congenital Methaemoglobinaemia who underwent caesarean section with successful outcome.

Case Report

A 20 yr old full term pregnant female weighing 60 Kg was referred to our centre with diagnosis of methaemoglobinemia. She had mild dyspnoea and bluish discoloration of nails since one month. There was no syncope, headache, palpitation or oedema. She gave history of cervical encerclage under spinal anaesthesia at 12 weeks. She did not have dyspnoea at that time.

On admission, she was conscious, oriented with RR of 18/min. She had a PR of 90/min and BP of 120/70 mm Hg. Systemic examination was normal. Her oxygen saturation was 86 % on air which increased to 90 % with oxygen. Her Hb was 14gm %, PCV was 45 % and methaemoglobin (MetHb) level was 24%. Her 2 D-echo and the foetal heart sounds were normal.

She was given Oxygen supplementation at 6-8 lit./min. Ascorbic acid 500 mg twice a day was started. Methylene blue was not indicated as her MetHb level was 24% (<30- 40%) [1]. Three days later, her MetHb level dropped to 19.5 gm % and oxygen saturation improved to 92-94 %. Arterial blood gas examination done at this time showed oxygen saturation of 93% at PaO2 of 200 mm Hg, $HCO3^-$ of 20.3 mmol/L, PCO2 of 32 mm Hg, and a pH of 7.43.

She was posted for caesarean section for post date pregnancy. Preoperative vital parameters were normal with oxygen saturation of 92-94%. Tablet Ranitidine 150 mg was given 2hrs before surgery. Metoclopramide was avoided. After preoxygenation, anaesthesia was induced with 250 mg Thiopentone, 60 mcg Fentanyl and 100 mg Succinyl choline. She was intubated with 6.5 no. cuffed endotracheal tube and maintained on oxygen + 1.7% Sevoflurane and intermittent Scoline. A healthy baby with apgar score of 9 was delivered. Intra operatively, her oxygen saturation varied between 94-96% with good cardiovascular stability. At the end of

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surgery, she was successfully extubated. Her postoperative course was uneventful. Her MetHb level was 17 gm % on 2nd postoperative day. Her G-6PD level was normal.

At the time of discharge, she was instructed to avoid contact with the room fresheners, well water, henna (mehandi), cosmetics, dyes, naphthalene balls, Primaquine, Dapsone and Sulfonamides [4-6] At follow up visit two months later, she had slight dyspnoea intermittently, without restriction of physical activity, fatigue or syncope. Her MetHb level was 10 gm %.

Discussion

Methaemoglobin is an oxygenation product of haemoglobin (Hb) where ferrous ion is oxidized to ferric form which is incapable of carrying oxygen [7]. Normally small amount of Hb is continuously being oxidized by endogenous agents. The level of MetHb stays below 1 gm % as it is continuously reduced by Nicotinamide Adenine Dinucleotide (NADH) b5 reductase enzyme present in RBCs. Hereditary deficiency of this enzyme causes congenital methaemoglobinaemia. NADH b5 reductase deficiency is a recessive trait. Only homozygotes or individuals who are compound heterozygotes express the disease. Heterozygotes are at increased risk of Methaemoglobinaemia when exposed to exogenous oxidative stress [8]. Patients of deficiency of NADH b5 reductase have normal survival and normal pregnancies as in our case [4] Their MetHb levels vary between 20-40%. Acquired methaemoglobinaemia which is relatively common than the congenital form also occurs in normal individuals exposed to drugs or chemicals that oxidize Hb at a rate that exceeds the rate of endogenous enzyme reduction [8].

The presence of MetHb above 1.5% causes cyanosis and is labeled as Methaemoglobinaemia. Levels up to 25-30 % do not cause symptoms except cyanosis and mild dyspnoea. Respiratory distress seen in mild degree of MetHb is because of left shift of oxygen dissociation curve of normal Hb units brought about by oxidized adjoining units [4]. This explains the dyspnoea in our patient. Levels above 30-40 % cause weakness, headache, tachycardia and giddiness. Lethargy, confusion, stupor and coma follow with levels of more than 50%. Circulatory collapse occurs at MetHb level more than 70 %. [6,9] ABG analysis is usually done to assess oxygenation status. In these patients, ABG does not give accurate idea of oxygen saturation as it is calculated from pH and pCO2 assuming Hb to be normal [9].

Acquired methaemoglobinaemia is suspected when sudden onset cyanosis is seen that is unresponsive to oxygen therapy and the CVS/RS are normal. Long standing symptoms or symptoms in siblings point to hereditary cause [4]. In our case, due to sub acute onset and absence of positive family history, we initially thought of an acquired aetiology. We could not, however, pinpoint the agent. In mild degree(<30%) of acquired form, once the causative agent is removed, MetHb levels come to normal within 48 hours even without treatment with reducing drugs [7]. In our case, it remained high (17%) in spite of treatment with ascorbic acid, which points towards congenital cause. The diagnosis of absence of enzyme NADH b5 reductase is done by enzyme essay but this facility was not available.

MetHb levels above 30 % need treatment. Methylene blue I.V. 1-2 mg/kg over 30min or orally 100-300mg/day is usually effective. Higher doses of methylene blue (>7 mg/kg) may cause haemolysis and persistent cyanosis, as the agent can paradoxically oxidize haemoglobin to MetHb. In patients with G-6PD deficiency, it is ineffective and can induce haemolysis. [6]. We found two reports of successful use of Methylene blue before C-section [1,2] Oral Ascorbic acid 500mg causes slower non enzymatic reduction of MetHb. Other treatment options are hyperbaric oxygen and exchange blood transfusion.

The drugs to be avoided in the peri operative period are local anaesthetics (benzocaine, prilocaine, lignocaine), nitroglycerine, nitroprusside, metoclopramide, ibuprofen, acetaminophen and nitrous oxide [5,10]. If procedure demands, local anaesthetics can be used in guarded doses keeping methylene blue ready. Intraoperative monitoring should include co-oximetry which detects and quantifies MetHb.

The MetHb level of the baby was normal. The baby was advised future follow up by a paediatrician as there are reports of neonatal jaundice, heart murmur, dyslalia and learning/memory impairement over the years in children born to mothers with Methaemoglobinaemia [11].

Conclusion

Monitoring by co- oximeter if available, keeping methylene blue ready, avoiding all causative factors and proper oxygenation form the pillars of successful anaesthetic management of a patient with Methaemoglobinaemia.

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