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NEONATAL CYANOSIS WITH DIARRHOEA – THINK BEYOND COMMON CAUSE

Neonatology

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ABSTRACT

Neonatal cyanosis is most commonly due to pulmonary and cardiac disease. Neonatal Methemoglobinemia is rare cause of Neonatal cyanosis. Methemoglobinemia not attributable to any drug or oxidant, has been described in neonate following gastroenteritis. Methemoglobinemia usually present with cyanosis,so always create diagnostic quandary. We report a case of methemoglobinemia in a neonate following diarrhea.

KEYWORDS

Cyanosis, Neonate, Methemoglobinemia, Diarrhea.

INTRODUCTION

Methemoglobinemia is rare cause of cyanosis. It is formed by oxidation of heme iron from ferrous(Fe++) to Ferric(Fe+++) form[1,2]. It constitutes less than 1% of total hemoglobin. Case report has been documented of methemoglobinemia following gastroenteritis with dehydration & metabolic acidosis[3]. Neonatal methemoglobinemia usually present with cyanosis and while investigating a case of Neonatal cyanosis, we usually think of cardiac and pulmonary causes that's why such scenario creates a diagnostic dilemma. So , Herewith we report a case of Neonatal methemoglobinemia following Diarrhea to stress on the fact that Methemoglobinemia should be suspected in case of gastroenteritis with cyanosis which is out of proportion to the history and clinical examination.

CASE REPORT

A 19 days old male baby brought in emergency with complaints of loose stool and vomiting for 1 day and bluish discoloration of body with respiratory distress so immediately baby shifted to NICU and electively intubated in view of respiratory distress and put on ventilator support.

His Birth history showed that he was a full term baby with birth weight of 3 kg. Antenatal period was uneventful and no history of maternal drug intake. Postnatally Baby was stable till day 19 of life, gaining weight with no history of drug intake or any surgical procedure.

On admission, baby was lethargic, dehydrated and cyanotic (central and peripheral cyanosis).Vital parameter were Temperature(36.5 C),Heart rate of 162/min, respiratory rate of 64/min,spo2 of 85% under room air, peripheral pulses feeble along with signs of dehydration.

On physical examination, Baby was lethargic and had labored breathing, bilateral air entry was present and equal and clear. Rest system were within normal limit. Initially baby was intubated and put on ventilator support . In view of dehydration, dehydration correction was given with intravenous fluid and intravenous antibiotic (Inj piptaz and amikacin) started. Despite on ventilator,spo2 not improved so Fio2 increased to 100% but still Fio2 remained below 89%.In view of presence of central cyanosis and poor saturation, To rule out CCHD,PPHN chest radiograph done showed normal lung field and cardiac shadow and 2D-ECHO revealed normal ventricular function with no structural anomaly.

Laboratory investigation – sepsis work up was within normal limit and blood culture was sterile.Serum electrolytes were normal. Stool routine microscopy was normal and culture was sterile.ABG analysis showed PH-7.40,P02 – 200 mm Hg,PCO2 – 35,HCO3-17.

BY ruling out cardiac and pulmonary causes, we think of possibility of rare form of hematological disorder so we performed Red Brown screening test for methhemoglobinemia was positive and we also review ABG for methemoglobin level which was very high approx. 60%.Hemoglobin electrophoresis done was normal and NADH cytochrome B5 reductase level was 5IU/g hemoglobin(Normal value is 10 – 200 IU/g).

After confirming the diagnosis, plan to give IV methylene blue so before starting Methylene Blue G6PD level should be determined because in G6PD deficient individuals, poor response to methylene blue is seen. So after confirming normal G6PD level baby was given Intravenous Methylene blue 2mg/kg. Methemoglobin level was drop down to 1.2% and baby showed clinical improvement in form of disappearance of cyanosis and gradually Spo2 level increased to 92-94% at Fio2 level of 21%.So,baby weaned off ventilator support after 24 hours. Baby dehydration was also improved after 48 hours so baby discharged after 3 days.

DISCUSSION

There are numerous common and rare causes of Neonatal cyanosis, One of the rare cause is Neonatal Methemoglobinemia so it should be suspected if newborn present with cyanosis and hypoxemia. After ruling out cardiac ,respiratory and infectious causes, if cyanosis did not improve with supplemental oxygen, Methemoglobinemia should be suspesected. In Methemoglobinemia, Heme iron is oxidized from (Fe++) to their ferric state(Fe+++).It act by shifting of oxygen dissociation curve to left[4] and unavailability of methemoglobin to transport oxygen that ultimately leads to decrease delivery of oxygen to tissue that leads to hypoxemia and acidosis.

Methemoglobinemia is of two types – Congenital and Acquired methemoglobinemia. Congenital methemoglobinemia is categorized into two main types-one due to Methemoglobin reductase enzyme(NADH cytochrome b5 reductase) and other due to Hemoglobin M[5]. Acquired methemoglobinemia occur because of disturbance in equilibrium between oxidation and reduction reaction due to exogenous factors like Nitro or amino derivative of benzene, derivatives of sulphur etc.

Methemoglobinemia can be associated in baby who is having diarrhea, metabolic acidosis or sepsis. Rationale behind this association is, in acidosis-activity of NADH methemoglobin reductase enzyme is disturbed that leads to decrease methemoglobin reduction.In septic baby,there is increased production and reduction of nitric oxide to nitrate which oxidizes the hemoglobin[6]. In diarrhea ,cause of methemoglobinemia is due to conversion of nitrates to nitrites.Another aspect is, Methemoglobin undergo reduction by cytochrome b5 reductase enzyme.It has been found that Neonatal level of cytochrome b5 reductase is much less than adult level[7] so they are prone to hemoglobin oidation and fetal hemoglobin is also more susceptible to oxidation.As in our case ,lower level of NADH cytochrome B5

reducatse level leads to methemoglobinemia and associated diarrhea and metabolic acidosis further increase the riSk of methemoglobinemia.

Methemoglobinemia is a potentially fatal condition in neonate required prompt diagnosis and treatment. So, Newborn with cyanosis and hypoxemia and cyanosis not improving with administration of supplemental oxygen,dark brown colored blood – Methemoglobinemia should be suspected and should be confirmed by Co-oximetry method,which is diagnostic test for methemoglobinemia. After confirmation of Methemoglobinemia – it should be etiological defined whether Congenital or Acquired.

Congenital methemoglobinemia diagnosed either by deficiency of cytochrome b5 reductase or Hemoglobin electrophoresis. Hemoglobin electrophoresis demonstrate Hemoglobin M,a hemoglobin variant causing cyanosis as a result of structural changes in alpha or beta chains. Deficiency of cytochrome b5 reductase is most common cause of congenital methemoglbinemia.

There is no specific treatment for congenital methemoglobinemia[8]. Treatment include avoid administration of oxidative compound and iv merthylene blue. IV methylene blue is indicated for asymptomatic patient with level greater than 30% and symptomatic patient with level > 20%.It has antioxidant properties,in presence of NADPH ,methylene blue is converted to leucomethylene blue result in non enzymatic reduction of methemoglobin[9].It shows prompt response in congenital methemoglobinemia due to deficiency of cytochrome B5 reductase as compared to HbM disease[10].If baby failed to show response to methylene blue – Hyperbaric oxygen and Exchange transfusion are alternative therapies [9,10].

CONCLUSION

Methemoglobinemia is life threatening disease in neonate if not diagnosed or treated promptly.So,it always be suspected if a newboprn present with cyanosis with hypoxemia,not improving with supplemental oxygen after excluding cardiac,respiratory and infectious cause.We also conclude that presence of lower NADH cytochrome B5 reductase level associated with diarrhea and metabolic acidosis increase risk of methemoglobinemia.

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