Acute glomerulonephritis secondary to G6PD deficiency with secondary Methemoglobinemia

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Abstract

Introduction: G6pd deficiency is a X linked chromosomal disorder affecting males and females are carriers. It presents with haemolytic anaemia and intravascular hemolysis. The enzyme deficiency causes decreased production of NADPH in the erythrocytes which keeps glutathione in reduced form preventing oxidative stress in cells. Secondary meth-haemoglobinemia occurs in susceptible individuals, due to decreased active form of haemoglobin available for carrying oxygen to cells. A 2 years old male child presented in the hospital emergency with complaints of cough, cold and fever for 3days and increased work of breathing with scanty brown coloured urine since 24hours. On examination there was severe pallor, mild perioral cyanosis and icterus. He had respiratory distress with bilateral crepitation, on auscultation the heart sounds are normal with S3 gallop rhythm. Child was managed conservatively and gradually improved with high concentration oxygen therapy and blood transfusion.

Discussion: the hemolysis was triggered by oxidative stress on the erythrocytes by acute infection and use of anti-pyretic drug. The child clinically improved with conservative treatment and was discharged.

Keywords: Acute glomerulonephritis secondary, g6pd deficiency, secondary Methemoglobinemia

Introduction:

G6PD deficiency affects around 400 million people worldwide and is characterized by considerable biochemical and molecular heterogeneity [8]. The prevalence of G6PD deficiency varied from 2.3 to 27.0 per cent with an overall prevalence of 7.7 per cent. Frequency of G6PD deficiency was observed in the Gamits (31.4%), Dhankas (20.4%), Warlis (19.6%), Dhodias (17.8%), Bhils (16.3%) and Garasiyas (15.2%). Bhils from Nasik district of Maharashtra have a very low frequency (1.4%). A complete absence of Gd gene was observed among the Mahadev Kolis from Ahmednagar district of Maharashtra [9]. G6PD deficiency is an X linked chromosomal disorder, which affects males and females are carriers. Glucose- 6 -phosphodiesterase is an important enzyme for maintenance of red blood cell energy production, it catalyses the first step of the hexose monophosphate shunt and gives one molecule of NADPH, which maintains the glutathione in reduced functional state, thus preventing oxidative stress. The most common manifestation is the episodic haemolytic anaemia caused by triggers (infection, use of drugs) and chronic non-spherocytic haemolytic anaemia.

Methemoglobinemia is a clinical syndrome caused by increased levels of methaemoglobin in serum as a result of congenital changes in the haemoglobin synthesis or metabolism leading to imbalance in reduction and oxidation of haemoglobin.

Case summary

A 2years male child presented in the hospital emergency with complaints cough, cold and fever since 3days with increased work of breathing and passage of scanty dark brown urine. On examination, the child was conscious and irritable. On general examination the child had severe pallor, mild perioral cyanosis, mild icterus. The pulse rate was 150/min, on auscultation normal heart sounds were heard with S3 gallop rhythm; Respiratory rate: 70/min with signs of respiratory distress and bilateral crepitation; blood pressure 90/50mmHg;
Discussion

G6pd deficiency occurs most commonly in the tropical and subtropical zones of eastern hemisphere in India. The prevalence is of 3% [2]. This was the first episode of haemolysis in this child, triggered by use of antipyretic drug for fever leading to oxidative stress on the red blood cells. Methaemoglobin is the oxidised form of haemoglobin, which does not bind to oxygen and increases affinity of O2 for the partially oxidised portion of haemoglobin and shifts the oxygen dissociation curve to the left [3]. The predominant pathway for methb reduction is NADPH dependent reaction catalysed by cytochrome b5, it utilises the NADPH produced by G6PD in hexose monophosphate pathway. The diagnosis depends on high degree of suspicion. These patients will not be maintaining the saturation in spite of normal pao2 and without significant cardiopulmonary insufficiency. Anaemia makes the patient more susceptible to Met-Haemoglobinemia by reducing the functional stores of Hb. Meth-haemoglobin levels above 10-15% change colour of the blood to brown and the child develops central cyanosis which is not responsive to O2 therapy [5]. Cardiovascular symptoms of anxiety, dyspnea and cyanosis are common between 20-30% which the patient had.

Treatment of Methemoglobinemia primarily depends on the severity of the disorder. Once the causative agent is removed the patient improves within a span of 36hours. Supportive therapy with supplemental oxygen increases plasma levels of dissolved oxygen. Methylen blue, an oxidising agent which is used in reduction of the meth-Hb levels but in g6pd deficiency the red blood cells do not produce enough NADPH to reduce the methylene blue to methylene leuco blue [8]. This patient responded to high concentration oxygen administration, two units of packed cell transfusion and supportive treatment.

In a case report by Marijin Schurman et al. noted the rare occurrence of Methemoglobinemia with g6pd deficiency after the boy ingested fava beans. The presentation was with anaemia and cyanosis and due to intravascular hemolysis developed acute renal failure.

In the a study done by Sarkar et al. [7] in children with intravascular haemolysis secondary to hemolysis due to G6pd deficiency found azotaemia in 20 patients. Those without oliguria were treated with supportive care and those with oliguria and acute renal failure were treated by dialysis. The principle intervention for reducing hemolysis in individuals with G6PD deficiency is avoiding exposure to drugs known to trigger hemolysis. In a 2016 systematic review of published reports, revealed no evidence of harm for vitamin C, vitamin E, vitamin K, ginkgo biloba, or alpha-lipoic acid when used in g6pd patients as supportive therapy [1].

References


