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Case Report

A 2-year-old boy with Glucose-6-Phosphate Dehydrogenase Deficiency Following Naphthalene Ingestion: a Case Report

*Ruhul A [1], Rashedul H [2], Shamima SS [3] Tanjia M [4]

Abstract:

Glucose-6-phosphate dehydrogenase (G6PD) deficiency is the most common hereditary enzyme disorder in the world and more than 400 million people have a deficiency in this enzyme. G6PD deficiency is an X-linked disorder, and one of its important signs is the presence of hemolytic anemia. It is a worldwide important cause of neonatal jaundice and causes life threatening hemolytic crisis in childhood. Here a 2-year- old boy was admitted into department of pediatrics of Sylhet Women's Medical College Hospital with fever for 2 days & passage of dark color urine for 1 day. There was history of accidental naphthalene ingestion present prior to develop symptoms. The boy was clinically febrile, severely pale, anicteric. No organomegaly was present. It was confirmed by G6PD level assay. Symptomatic & supportive treatment was given with packed cell volume transfusion and avoidance of triggering factors was advised. Genetic counseling was also done.

Keywords: Hemolytic anemia, Glucose-6-Phosphate Dehydrogenase Deficiency

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Introduction:

Glucose-6-phosphate dehydrogenase deficiency is the important enzyme deficiency worldwide. Glucose-6-phosphate dehydrogenase is the rate limiting enzyme in the pentose phosphate pathway, which generates NADPH, required for the production of reduced glutathione in RBCs, and it is essential for the function of catalase. These provide cellular protection from oxidative stress. G6PD activity decreases significantly with aging of RBCs over time. A report in 1976 stated that several drugs were responsible for hemolytic effects in G6PD-deficient individuals.[1]

1. Dr. Ruhul Amin, Assistant Professor, Dept. of Paediatrics, Sylhet Women's Medical College Hospital.
2. Dr.Rashedul Haque, Professor, Dept. of Paediatrics, Sylhet Women's Medical College Hospital.
3. Dr. Shamima Sharmin Shova, Assistant Professor, Dept. of Paediatrics, Anwar Khan Modern Medical College Hospital
4. Dr. Tanjia Masuma, Medical officer, Dept. of Paediatrics, Sylhet Women's Medical College Hospital

Corresponding author: Dr. Ruhul Amin,
Assistant Professor, Dept. of Paediatrics, Sylhet Women's Medical College Hospital.
Email: ruhulrumi37@gmail.com

Now a days, G-6PD deficiency is the most common hereditary enzyme deficiency, causing hemolysis. Glucose-6-phosphate dehydrogenase (G6PD) deficiency affects more than 400 million people around the world, representing an overall 4.9% global prevalence.[2,3] The majority of individuals with Glucose-6-phosphate dehydrogenase deficiency are asymptomatic although there is a risk of neonatal hyperbilirubinemia and acute hemolysis after contact with oxidative stress.[4] Frequency of G6PD is relatively high among African, Americans in the United States (13%) and populations in the Mediterranean (5 to 40%). G6PD deficiency is an X-linked enzyme defect, and one of its important signs is the presence of hemolytic anemia. Hemolysis may be triggered by infections, drugs with oxidative properties, such as acetyl salicylic acid, vitamin K, chloramphenicol and antimalarial drugs.[3,5] So advice is given to avoid cross contaminations, fava beans as acute crisis episodes have been reported following fava beans DNA.[6] The gene coding for G6PD is located on the X chromosome; therefore, transmission is hereditary and sex-linked.[7] G6PD deficiency is caused by mutations located at Xq28, consisting of 13 exons and 12 introns as G6PD gene. The G6PD deficiency is an X-linked recessive defect that exhibits several clinical manifestations as

jaundice, hemolytic anemia, splenomegaly, and hemoglobinuria.[8] The present study describes the case of a patient with G6PD deficiency with urinary tract infection.

Case Report:

A 2-year-old boy was admitted in the department of pediatrics of Sylhet Women's Medical College Hospital, Sylhet with the complaints of fever for 2 days which was low grade, intermittent in nature, not associated with chills and rigor and subsided by taking antipyretic. Mother also complained of passage of dark color urine for 1 day. He had no history of jaundice, abdominal pain, convulsion or unconsciousness. He is the 3rd issue of his non-consanguineous parents. There was history of accidental naphthalene ingestion present prior to develop of these symptoms. Clinically he was ill looking, severely pale, anicteric, febrile, pulse 96 b/min, R/R-28br/min. Abdomen was soft, non-tender without any organomegaly. The Laboratory findings revealed Hb 2.8 gm/dl, TC of WBC 24000/cm, HCT 8.7%. PBF showed picture of hemolytic anemia. Routine examination of urine showed pus cell 10-15/HPF, no RBC was present. Our working diagnosis was G6PD deficiency with urinary tract infection and the patient was treated immediately with transfusion of packed cell volume & antibiotic. Post transfusion Hb was 10.5gm/dl. After giving immediate management, patient was improved and we advised the patient to do G6PD assay after few weeks and report showed 5.13 u/gram Hb (8.5-13.5). So the final diagnosis was G6PD deficiency with urinary tract infection.

Figure 1. Dark urine

Table 1. Laboratory examinations

Parameters	Result	Reference Range
Hemoglobin	**Hb 2.8 gm/dl**	**10.7-14.7 g/dl**
Erythrocyte	0.88 x 10^6	4.1-5.3x106 /mm3
HCT	8.7%	37-54%
Leucocyte	24 x 10^3	4-11 x 103/mm3
Platelet	366 x 10^3	150-400 x 103/mm3
G6PD level	**5.13 u/gram Hb**	**8.5-13.5 u/gram Hb**

Discussion: Glucose-6-phosphate dehydrogenase deficiency is one the commonest hereditary enzyme deficiency in the world. The majority of Patients are asymptomatic. Acute hemolysis occurs after contact with oxidative stressors, infection, chemicals and oxidant drugs. Our patient presented in the emergency room with feature of severe hemolysis. We suspected hemolytic anemia based on feature of hemolysis and history of naphthalene exposure. Diagnosis was confirmed by enzyme assay. The causative factor of hemolysis of our presented case was naphthalene. Occurrence of acute Hemolysis due to exposure of naphthalene in G6PD individuals have been reported worldwide.[9] Among the Bangladeshi child, toxicity of naphthalene is not very commonly reported. Rahman et al. reported a 22 years woman having hemolytic anemia after accidental ingestion of liquid substance containing naphthalene at 2012 for the first time in Bangladesh. In that case the woman had normal level of G6PD .[10] In present reported case, acute Hemolysis occurred in a 2 year old Bangladeshi G6PD deficient child after exposure of naphthalene. Naphthalene is well absorbed by oral exposure, it can be also absorbed through the skin and inhalation.[9] In the present case, the child used to play with naphthalene, so dermal absorption was taking place to the child for last few days. As the child was G6PD deficient, he developed severe episodes of hemolysis even after ingesting very small part of a naphthalene. Here, naphthalene played a triggering role as an

oxidant causing hemolysis. Ingestion of naphthalene causes production of excessive free oxygen radicals resulting lipid peroxidation and damage to deoxyribonucleic acid .[9] It is well known that, G6PD deficient patients have low tolerance to oxidative free radicals .[11] Previous different studies, also showed that naphthalene is a possible triggering agent of acute Hemolysis in previously well child and G6PD deficient individuals .[12]

Conclusion: G6PD deficiency usually presents with pallor, jaundice and dark urine after exposure to precipitating factors as like as naphthalene. Naphthalene is a threat for G6PD deficient child and normal individuals. So it should be avoided for household use.

Conflict of interest: No conflict of interest

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Ethics approval: Informed consent was obtained from the patient parents before the case report was written

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