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Case Report

A Case Report of a Teaspoon Portion of Fava Beans Caused Severe Hemolysis in G6PD-Deficient Baby: Time to Include G6PD Enzyme Testing in the Premarital Screening Program and Newborns

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Keywords
Fava beans · G6PD · Hemolysis · Premarital screening

Abstract
A 2-year-old baby of Arabian descent presented to the emergency department (ED) with signs of acute hemolytic anemia due to consuming a teaspoon portion of fava beans. Besides jaundice, nausea, vomiting, and fatigue, dark urine was the primary motive for the ED visit. The baby was diagnosed with glucose-6-phosphate dehydrogenase (G6PD) deficiency, which was adequately managed. Upon discharge, he was furnished with a nutritional and medical counseling plan to avoid the triggering factors of hemolysis. Hospital management of this and other cases imposes a high cost on health systems. One of the cost-effective measures to reduce that cost on health systems is to incorporate the G6PD enzyme test in newborns or premarital screening programs. Thus, the proposed measures would have eliminated the suffering of these innocent souls.

Introduction

Glucose-6-phosphate dehydrogenase (G6PD) deficiency is the most common enzyme deficiency syndrome globally, and it is an X-linked genetic disorder which predominantly affects males. Essentially, G6PD protects red blood cells against oxidative damage by

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converting nicotinamide adenine dinucleotide (NAD) + hydrogen (H) (NADH) to NADPH and destroying reactive oxygen species and free radicals by maintaining glutathione in its reduced form [1]. G6PD deficiency makes red blood cells vulnerable to oxidative stress, breakdown, and thus hemolytic anemia. Certain factors were reported triggering hemolysis, including fava bean consumption, infections, anti-malaria drugs, and some antibiotics such as nitrofurantoin and sulfa [1]. Financially, G6PD deficiency hospital management enforces a tremendous cost on the health system, which needs evaluation of other cost-effective measures. In this instance, we report a case of a two-year-old child presented with G6PD deficiency-related hemolysis secondary to ingestion of a teaspoon portion of fava beans that could have been avoided if this child had already been tested for G6PD at the time of berth or if his parent's premarital screening included G6PD deficiency test as his mother stated.

Case Presentation

A 2-year-old baby with no medical or family history of anemia presented to the emergency department (ED) with 3 days of fever (39.1°C), vomiting, nausea, pallor, jaundice, and unusual dark color of the wet diaper for 1 day. His mother stated that her baby experienced these symptoms 3 days after eating a small portion of cooked beans. His labs revealed reticulocytosis, hemoglobin of 5.1 g/dL, total serum bilirubin of 68.1 μmol/L, serum direct bilirubin of 5.0 μmol/L, and indirect serum bilirubin of 63.1 μmol/L. Examination of the urine sample revealed dark, no albumin, no glucose, no acetone, no pus or red blood cells, and +++ urobilinogen. Hemoglobin electrophoresis revealed a typical pattern (A_1 = 96.8%, F = 0.3%, and A_2 = 2.9%). Upon admission, the patient was administered cefotaxime 500 mg IV every 8 h and an adol suppository 100 mg every 4 h. In addition, he transfused 150 mL of the same group of fresh-packed red blood cells. The next day, his hemoglobin increased to 8.1 g/dL. The baby was diagnosed with acute hemolytic anemia due to G6PD deficiency after his G6PD test showed a low level (2.0 U/g Hb) correlated with the clinical history and features. After 2 days of admission, the patient was discharged with 8.9 g/dL hemoglobin and clear urine. He has been prescribed folic acid 1 mg and multivitamins with a nutritional counseling sheet for what to avoid.

Discussion

G6PD deficiency is one of the world's most common hereditary blood disorders, especially in the Mediterranean, Middle Eastern, and sub-Saharan Africans and East Asians [2]. The hemolytic risk factor in this patient was identified as fava bean consumption, though it was a teaspoon portion. It was reported that a 4-year-old patient, ingesting 40–60 gm of fava beans caused severe hemolysis, proving that even a teaspoon of fava beans can be harmful to G6PD-deficient patients [3]. Though this patient was transfused with 150 mL of fresh-packed red blood cells, vitamin supplements, mainly folate, and vitamin E play an essential role in reducing the symptoms of G6PD deficiency in this patient. Iron, folate, and multivitamin supplements are valuable treatments for G6PD patients where they can reduce oxidative-induced hemolysis and maintain excellent oxidative stress in G6PD deficiency patients.

Upon hospital discharge, parents were provided with a nutritional management plan for their child's diet to avoid the triggering factors of hemolysis. The plan advised on the exclusion of fava beans and avoidance of any bean derivatives or contaminated food. In addition, hydration is strongly recommended to reduce oxidative stress and muscular fatigue. Eating

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food rich in antioxidants, like dates, grapes, spinach, oranges, and berries, was also advised to minimize the risk of free radicals and thus prevent hemolysis. A list of certain drugs, which trigger hemolysis, is also provided. The nutritional management plan is the primary technique to prevent hemolysis and oxidative stress in G6PD deficiency patients.

In addition to a proper nutritional counseling plan, G6PD deficiency patients and their guardians are always encouraged to understand and recognize the symptoms of acute hemolysis and seek urgent medical treatment when required. In this case, the child had been suffering from fever and hemolysis symptoms for 3 days before attending the ED. His mother reported that the observation of dark urine was her main motive for the ED visit. Unmanaged or untreated G6PD deficiency, especially in children, might lead to acute hemolytic anemia with life-threatening complications, including shortness of breath, rapid heart rate, kidney failure, and brain damage.

Even though there is no statistical data on G6PD deficiency cases in the Kingdom of Saudi Arabia, hemolytic anemia due to G6PD deficiency still counts as one of the leading causes of red blood cell transfusion in hospitals, especially in the regions' where arranged marriage is still practiced [4]. Premarital screening for certain hereditary hemoglobin disorders (sickle cell anemia and thalassemia) and genetic counseling programs remarkably influence reducing these diseases [4]. However, the Saudi premarital screening and the genetic counseling program do not include G6PD deficiency testing, in which cases of G6PD deficiency cannot be reduced.

Further studies should be evaluated after the cost of the screening versus the cost of the G6PD deficiency treatment is promptly considered. In their study of the comparison between the medical expenses of G6PD deficiency treatment and G6PD enzyme screening, Derbandi et al. [5] stated that the estimated cost of the treatment of G6PD deficiency hospitalized patients (Rasht City, Iran) was higher than the cost of the G6PD enzyme screening for all the newborn infants in the city between October 2011 and September 2012.

Moreover, Khneisser et al. [6] proved the efficiency of the routine testing for the G6PD enzyme at a newborn level in Lebanon. They found out that the risk of hospitalization due to the crisis had decreased by 95% among those patients tested for G6PD deficiency. Both studies proved the positive impact of the G6PD enzyme test, and it is a cost-effective potential on health systems [5, 6].

Another cost-effective potential is premarital screening for the G6PD enzyme. Including the G6PD enzyme in the premarital screening program would reduce the number of new G6PD deficiency cases. The partners might consider cancellation of their marriage if they received the proper counseling when one of them has the mutated gene. They would also have a clear vision of this disease and be able to manage such cases if they had G6PD-deficient offspring. Consequently, the new positive cases and the cost imposed on the health systems would decrease.

Conclusion

G6PD deficiency is one of the world's most common blood hereditary disorders and one of the leading causes of blood transfusion in Saudi Arabia. In addition to avoidance of the triggering factors of hemolysis, G6PD enzyme screening programs should be adopted to reduce the prevalence of this genetic blood disorder. Testing for the G6PD enzyme at birth or including it in the premarital screening is one of these programs. It would quickly end the suffering of the new G6PD patients and lower the cost imposed on the healthcare systems. The CARE Checklist has been completed by the authors for this case report and is attached as supplementary material.

Statement of Ethics

Ethical approval is not required for this study in accordance with local or national guidelines. A written informed consent was obtained from the parent/legal guardian of the patient for publication of the details of their medical case and any accompanying images.

Conflict of Interest Statement

The authors further declare that the study holds no conflicts of interest.

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Author Contributions

Ahmad Alqarni's contributions were in drafting the work and revising it critically for important intellectual content. Ahmed Hjazi's contributions were in writing the final draft and submitting the paper.

Data Availability Statement

All data generated or analyzed during this study are included in this article and are not publicly available due to the Saudi Health governmental restriction of patient confidence but are available from the corresponding author upon reasonable request.

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