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CASE REPORT – OPEN ACCESS

Therapeutic Plasma Exchange in G6PD Deficient Patient Complicated with Intravascular Hemolysis: A Case Report

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ABSTRACT

Glucose-6-phosphate dehydrogenase is an enzyme that protects the erythrocytes against oxidative damage. G6PD deficiency is a common disorder in Gulf Countries that may present with hemolytic anemia and acute renal failure after exposure to oxidative triggers, such as drug, infection or some foodstuffs. In this study, a severe and complicated case of a G6PD deficient patient was presented. A 65-year old G6PD deficient patient presented with acute renal failure and intravascular hemolysis after eating fava beans. Therapeutic plasma exchange was used in the patient for two consecutive days. Clinical and laboratory findings were improved with plasma exchange. Although supportive therapy is enough for mild cases, therapeutic plasma exchange can be life-saving in complicated cases.

Keywords: Plasma exchange, free hemoglobin, G6PD, kidney injury, hemolysis

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INTRODUCTION

Glucose-6-phosphate dehydrogenase (G6PD) deficiency is the most common enzyme deficiency of red blood cells. It is an X-linked disorder, which is common in the Mediterranean, African and some East Asian populations. Diagnosis is based on symptoms and supported by blood tests and genetic testing (1). This deficiency makes erythrocytes sensitive to oxidative stress that can be triggered by some factors, such as toxins or drugs, and this causes hemolysis. Severe intravascular hemolysis may cause renal damage with an increased risk of morbidity and mortality (2).

In here, an outcome of a G6PD deficient patient complicated with severe intravascular hemolysis and acute renal failure due to eating fava beans was treated with plasma exchange and presented.

CASE REPORT

A 65-year old male with known G6PD deficiency presented at the Emergency Department with complaints of dark urine and shortness of breath. The medical history revealed dark red urine, which had gradually had worsened over the past three days after eating a large amount of fava beans. Two days after eating the fava beans, he also developed mild dyspnea, which worsened on admission day.

The general physical examination determined marked pallor, jaundice, tachycardia and tachypnea. The spleen and liver were normal in size. Complete blood count showed the following; Hemoglobin (Hb) 9.7 g/dl, Hematocrit 26.8%, Mean Corpuscular Volume (MCV) 92 fl, Mean Corpuscular Hb (MCHb) 33 pg, white blood cell count $18.1x10^9$/L and platelet count $185x10^9$/L.

The biochemistry test results indicated intravascular hemolysis with the following; indirect bilirubin 1.8 mg/dl (less than 0.8), LDH 2498 U/L (100–190 U/L), Serum Creatinine 1.6 mg/dl (0.6–1.2) and corrected Reticulocyte Count 5 % (0.2–2.5%). His potassium level was in normal limit, and blood PH was 7.30.

Peripheral smear (Wright Giemsa stain, x 400) showed: normocytic normochromic RBCs with anisopoikilocytosis (many blister cells, occasional bite cells and some RBCs showed Heinz bodies inclusions), moderate polymorph nuclear leukocytosis with absolute lymphopenia, and the platelet count was normal with some large platelets seen (Fig. 1). G6PD level was low at 0.3 U/g Hb (normal range: 10–10.4).

Intravenous normal saline was started as supportive treatment. At less than 24 hours after admission, the hemoglobin level of the patient dropped to 6.7 g/dl with increasing creatinine and LDH levels, as seen in Table 1.

Therapeutic plasma exchange (TPE) procedures were performed on two consecutive days with fresh frozen plasma using a Spectra Optia® machine. Acid citrate dextrose solution A (ACD-A) was used as an anticoagulant. The total plasma volume was changed 1.2 fold. Three-units pocket of red blood cells were given after the plasma exchange.

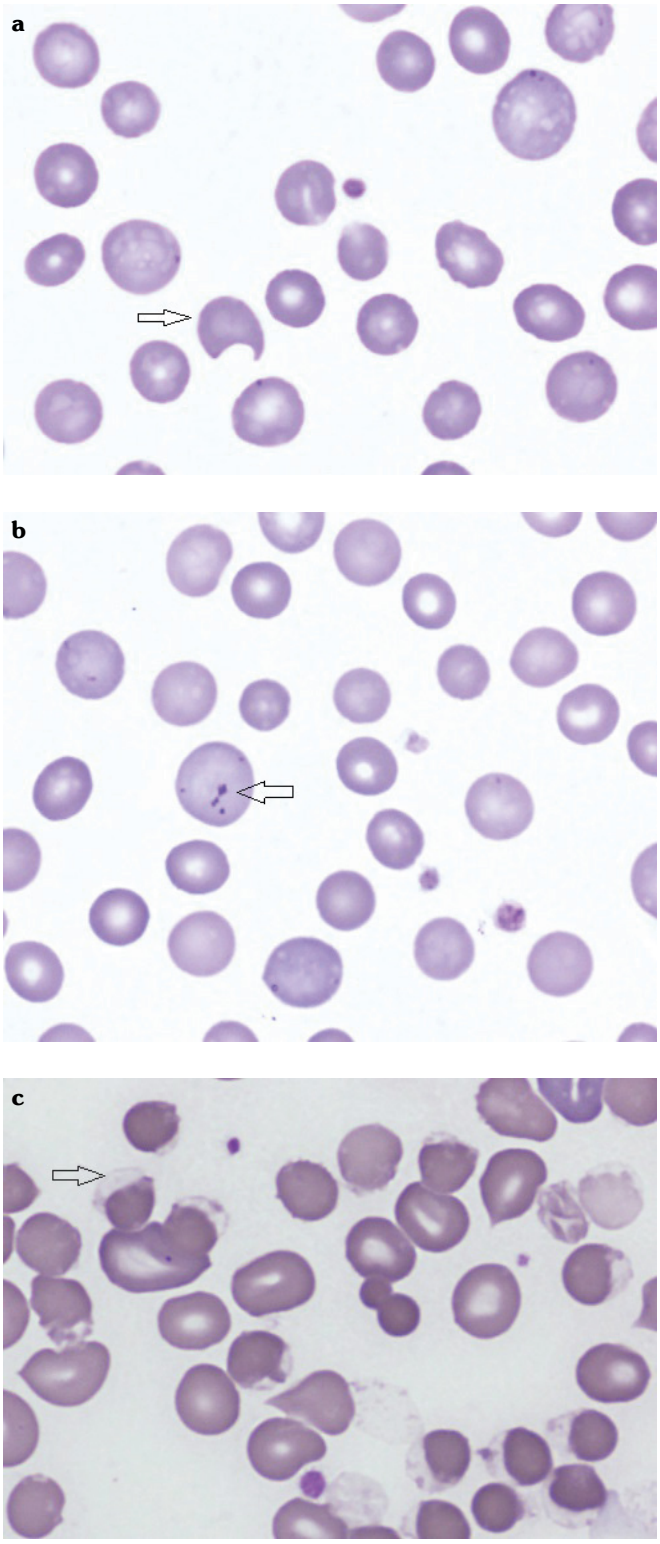

Figure 1. **(a) Peripheral blood smear shows bite cell. (b) Peripheral blood smear shows Heinz bodies. (c) Peripheral blood smear shows many blister cells**

Shortly after completion of TPE, the hemolysis parameters and renal function began to improve. The patient was discharged one week after admission. Patient consent was obtained for this study.

Table 1. Laboratory findings before and after therapeutic plasma exchange

Parameters	Follow up				
	12 hr. before TPE	12 hr. after TPE	24 hr. after TPE	2 weeks after TPE	4 weeks after TPE
Hb (gr/dl)	6.7	9.7	10.4	12.2	14.7
LDH (U)	3199	784	524	180	160
Bilirubin total (mg/dl)	10.6	46.8	10.7	–	10.5
Bilirubin indirect (mg/dl)	10	39	6.77	–	8.2
Creatinine (mg/dl)	1.8	2.1	1.9	1.5	1.3
Reticulocyte rate %	29	15.3	–	–	2.9

TPE: Therapeutic plasma exchange; Hb: Hemoglobin; LDH: Lactate dehydrogenase

DISCUSSION

G6PD deficiency, a rare disorder worldwide, is a common X-linked genetic disease with a high prevalence of up to 30% in Arab countries (1, 3). This deficiency may cause acute hemolysis due to a lack of antioxidant protection in the setting of oxidant injury by drugs (e.g., Primaquine, Sulfanilamide, Isobutyl nitrite), acute diseases, and certain foods, such as fava beans (4). Hemolysis is generally seen as mild and self- limiting. Supportive therapy is sufficient in most cases. The physicians should avoid drugs or factors that precipitated disease. Fava beans precipitated intravascular hemolysis in our case.

Renal damage can be seen due to severe hemolysis as a result of tubular damage that arises from free hemoglobin. This result may cause progressive renal failure. For the reason of high mortality and complication in these cases, more intensive treatment is needed (5). Kidney injury that arises from free hemoglobin is commonly managed with hemodialysis. TPE is an extracorporeal treatment that effectively removes antibodies, toxins, and immune complexes from the plasma and replaces it with another physiological fluid. TPE is to remove free hemoglobin substances from the plasma to allow the resolution of the disease and/or to decrease morbidity (5–7). TPE is also successfully used in patients having hemolytic anemia, regardless of immune or non-immune originated (8). However, only two cases have been reported in the literature, who have G6PD-deficiency (9, 10). Hemodialysis was not used in this case because the patient had no acidosis and electrolyte imbalance. After TPE, LDH, indirect bilirubin tests were returned to the normal limits in a short time.

CONCLUSION

Plasmapheresis might be considered for the treatment of G6PD deficiency-related severe intravascular hemolysis. Clinicians working in G6PD endemic areas should be aware TPE treatment option for the management of severe intravascular hemolysis.

Informed Consent: Written informed consent was obtained from patients who participated in this study.

Peer-review: Externally peer-reviewed.

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Conflict of Interest: There is no conflict of interest in this study.

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