Methemoglobinemia and Intravascular Hemolysis in a Patient with G6PD Deficiency

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Dear Sir,

We report the observation of a patient on chronic hemodialysis who presented with acute hemolytic anemia and methemoglobinemia. An etiologic investigation carried out confirmed G6PD deficiency.

Ms. F., of white origin, on chronic hemodialysis since 1985 for hepatorenal polycystic disease, presented with a slate-colored peripheral cyanosis and hypoesthesia 12 h after hemodialysis. Blood pressure and auscultation of the thorax were normal. The sample of blood was chocolate-brown colored, blood gas showed a compensated metabolic acido-sis, oxygen saturation was 99.2%. Hemolytic anemia was severe: hemoglobin 5.5 g/dl and haptoglobin 50 g/l. Diagnosis of methemoglobinemia was suspected and confirmed retrospectively: 17.5%. An injection of vitamin C 500 mg was inefficient; on the other hand, cyanosis disappeared 1 h after the infusion of methylene blue 1 mg/kg (methemoglobinemia: 9.4%). However, her neurological state and hemolysis worsened, requiring massive transfusions. The clinical state was progressively ameliorated, methemoglobinemia became normal at day 8 and hemolysis disappeared at day 10. Etiologic investigation was carried out. The patient had taken a paracetamol tablet on her previous dialysis session. Hemoglobin electrophoresis was normal, investigations for infectious diseases were negative. The water treatment is classic, particularly with micropore filters, a water softener, an activated carbon and a reverse osmosis module. The day after bacteriological and chemical analysis of the supplied water had been carried out, chloramines and nitrites were not detected, nitrate concentration was 1.19 mg/l. We have learnt afterwards that the hospital had been temporarily supplied with pit-water the day the incident occurred. G6PD activity measured 2 months after the transfusions in our patient revealed a partial G6PD deficiency: 50 U/10¹² RBCs (normal range 140-370). The patient's son, also victim of a hepatorenal polycystic disease with moderate renal failure and anemia of 9 g/dl, was equally deficient in G6PD.

Methemoglobinemias associated with hemodialysis are classically attributed to an elevated concentration of chloramines or nitrates in the distributed water [1]. The search of an eventual G6PD deficiency capable of provoking methemoglobinemia by inhibition of NADPH-flavine reductase should always be carried out in a case of methemoglobinemia associated with

intravascular hemolysis [2]. Symptomatic treatment in the absence of G6PD deficiency should be methylene blue. The latter, in the presence of NADPH, is reduced to leukomethylene blue and may, without intermediate, reduce methemoglobin. In case of G6PD deficiency, methylene blue is inefficient and may on the contrary exacerbate hemolysis [3].

In our patient with a partial deficiency of G6PD, it is likely that the presence of nitrates has acted as an oxidizing agent. In fact, the

pit-water supplied on the day of the incident is well-known for its richness in nitrates. A synergic action with paracetamol should not be totally excluded, even though consumed frequently before. Methylene blue injection might have aggravated intravascular hemolysis, the disappearance of cyanosis can be attributed to a decline in reduced hemoglobin.

Every methemoglobinemia in the course of hemodialysis then justifies a careful bacteriological, chemical and erythrocyte study. G6PD deficiency affects more than 200 million people worldwide, so dosage of this erythroenzymopathy activity should be realized systematically.

References

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