Use of thromboelastography and hydro-electrolytic management in a child with chronic renal insufficiency submitted to liver transplantation secondary to type 1 primary hyperoxaluria


Sir—Type 1 primary hyperoxaluria (PH1) is an innate error of the metabolism characterized by the deficiency or absence of the peroxisomal hepatic enzyme alanine-glyoxalate aminotransferase, which promotes the transamination of glyoxalate to glycine. Renal and skeletal systems are the main targets of the disease that also may involve heart, nerves, joints, arteries, skin, soft tissues, and retina (1). No form of dialysis is capable of overcoming oxalate production, and isolated kidney transplantation is unable to prevent the progression of the disease. A better understanding of PH1 has led to the introduction of liver transplantation (LT) as a form of gene and enzyme replacement (2). A LT on a nephropathic patient can lead to unpredictable and accentuated problems considering blood coagulation and hydroelectrolytic management.

A 13-year-old girl, 25 kg, diagnosed with oxalosis, already submitted twice to renal transplantation, evolving with progressive loss of renal function was sent to perform an orthotopic LT. The patient was monitored with EKG (HR: 124 b-min⁻¹), noninvasive and invasive arterial blood pressure (MAP: 90 mmHg), pulse oximetry (SatO₂: 98%), central venous pressure (CVP), nasopharyngeal thermometer (36.6°C/176°C), vesical probe and serial examination collections were performed. Coagulation was monitored by means of serial thromboelastography (TEG).

The patient maintained hemodynamic stability, without the need for vasoactive amines in continuous infusion. Normal filling pressure was maintained (CVP 6–8 mmHg). Volemic replacement was performed with a solution of Ringer’s lactate with albumin 7% in a total of 1350 ml and 300 ml of erythrocyte concentrate. Just prior to reperfusion, 10% calcium chloride (0.2 ml·kg⁻¹), insulin (0.1 UI·kg⁻¹) and 10% glucose (0.4 ml·kg⁻¹) were administered. Upon reperfusion, two boluses of epinephrine (10 µg each) were administered because of a drop in arterial pressure associated with a reduction in heart rate. Following reperfusion, the patient maintained hemodynamic stability again, with signs of the functioning of the hepatic graft.

TEG curves prior to the surgical incision revealed normal coagulation (not shown), which was maintained through the anhepatic phase. After reperfusion, heparinoids were detected by TEG (Figure 1), simultaneously with diffuse bleeding in the operating field. Protamine (25 mg) was administered, resolving the bleeding and allowing the closure of the surgical incision without excessive drainage through the drain. The patient was sent to the PICU intubated and hemodynamically stable. She evolved with no need of vasoactive amines or blood transfusion, was extubated 12 h after the end of the surgery and discharged 36 h later from the PICU.

Oxalosis is a hepatic disease with extra-hepatic complications, presenting with kidney failure and maintaining normal hepatic synthetic function. It accounts for 1% of LT causes (3). Considering fluids balance, to avoid hypervolemia, hypercloremic acidosis, excessive edema, poor distribution of volume and compromised anastomosis, infusion was performed with Ringer’s lactate and albumin (7% final concentration of albumin). The objective was to maintain oncotolic pressure and to replace the losses, with lesser extravasations to the interstice with a lower total volume administered.

The prevention of hyperkalemia is essential in a patient with previous chronic renal insufficiency (CRI). Thus, before reperfusion, insulin and calcium chloride were administrated. Insulin is the fastest and most consistent agent for reducing the concentration of serum potassium and its mechanism of action is independent of its action on the metabolism of glucose. Maximal hypokalemic response occurs at a serum insulin concentration of about 1/5 of that necessary to achieve the maximal hypoglycemic response. The capture of potassium occurs within seconds.
after the administering of insulin, unlike the glucose response, which requires 30–60 min. The marked resistance of the glycemic metabolism to insulin makes the co-administering of glucose unnecessary and it is better to monitor glycemia at 30-min intervals (4). Here, the greater need for glucose after reperfusion, even with good signs of the functioning of the graft was attributed to the longer plasma half-life of the insulin because of CRI.

TEG measures the viscoelastic and mechanical properties of the developing clot. TEG allows a differential diagnosis of the coagulopathy and separation of surgical and non-surgical bleeding. It allows in vitro tests, such as therapeutic trials, but is unable to identify the specific factor in deficiency (5). During LT, heparinoids may appear after reperfusion of the graft and normally exhibit a self-limited behavior because of their rapid elimination. The inclusion of protamine in the TEG examination removes the heparin-like effect from the parameters of the sample tested, serving as a therapeutic trial (6). The presence of heparinoids was attributed to its longer elimination half-life associated to renal insufficiency. Protamine was administered to counteract the heparin-like action of the heparinoids, with a clinical improvement in bleeding. The aim was not the normalization of coagulation, but to avoid bleeding while the cause (heparinoids) was eliminated by the organism. With TEG, the infusion of blood products was guided, avoiding hypervolemia, trombosis of the anastomoses and the exposition of the patient to all risks associated to transfusions.

In our case, a meticulous anesthesia technique, considering all the particularities of the patient, resulted in a successful transplant with no need for postoperative blood transfusions, re-operation because of bleeding or vascular stability until cardiopulmonary bypass could be established. Ventilation gradually improved and the surgery proceeded uneventfully. The child was successfully weaned off bypass and transferred to cardiac intensive care for further management.

Basiliximab is a chimeric monoclonal antibody that specifically binds to the alpha chain of IL-2 receptors; therefore, blocking IL-2 mediated activation and proliferation of T-lymphocytes. It is being increasingly used in heart transplantation where it has been shown to be safe, with no increased incidence of infection (1,2). A recent

References

Acute hypersensitivity reaction on re-exposure to basiliximab in an infant undergoing heart transplantation


Sr—We would like to report a case of a severe hypersensitivity reaction to basiliximab in a 7-month-old female infant, who presented for heart transplantation at our institution. She had been diagnosed with dilated cardiomyopathy (Fractional shortening = 6.7%) at 7 weeks of age. This was her second presentation for cardiac transplant; she had been scheduled for cardiac transplant 2 months previously but the procedure had been cancelled because of donor complications. She had received basiliximab as part of her induction immunosupression on the first presentation, immediately prior to cancellation of the procedure. On this occasion, induction of anesthesia with ketamine 1 mg·kg⁻¹ and fentanyl 3 μg·kg⁻¹ was uneventful. A nasotracheal tube was inserted after pancuronium 0.2 mg·kg⁻¹. Arterial and central venous access was then secured whilst the transplant nurse coordinator administered antibiotics (cefazidime and teicoplanin), followed by an intravenous injection of basiliximab 10 mg, as per protocol.

A few minutes after the basiliximab was given, the infant was noted to have a widespread rash with bilateral wheezing on auscultation and reduced tidal volumes. Cardiovascular stability was initially maintained and hydrocortisone 4 mg·kg⁻¹ was administered. Severe hypotension and bradycardia followed, requiring immediate advanced life support. A fluid bolus of 10 ml·kg⁻¹ Hartmann’s solution and 10 μg·kg⁻¹ of intravenous epinephrine were given. An epinephrine infusion was commenced and continued at 0.1 μg·kg⁻¹·min⁻¹ to maintain cardiovascular stability until cardiopulmonary bypass could be established. Ventilation gradually improved and the surgery proceeded uneventfully. The child was successfully weaned off bypass and transferred to cardiac intensive care for further management.

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