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Case of the week 03/2022

Hemoadsorption as bridge to liver transplant in a six-month old patient with hepatic failure

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Journal of Pediatrics and Neonatology 2021; 2:1017

This case reports on a 6-month-old female patient (8 kg, 55 cm), who was admitted to the hospital due to severe hyperbilirubinemia.

Case presentation

- The patient was a known case of Progressive Familial Intrahepatic Cholestasis (PFIC) undergoing genetic work-up
- On admission, she was awake and breathing spontaneously
- Continuously measured mean arterial pressure (MAP) was 60 mmHg, heart rate 110/min, and SpO_2 100%
- She exhibited signs of jaundice and a painful abdomen, indicative of acute on chronic liver failure (AoCLF)
- Laboratory data showed a white blood cell (WBC) count of 23,520/mm^3; platelets (PLT) 231,000/ mm^3; hemoglobin (Hb) 11 g/dl; C-reactive protein (CRP) 5.6 ng/l; lipase 175 U/L; amylase 3.04 U/L; ammonia 219 µg/dl; lactate dehydrogenase (LDH) 381 mU/ml; total bilirubin 29.42 mg/dl (indirect 9.36 mg/dl, direct 20.06 mg/dl); creatinine phosphokinase (CPK) 252 mU/ml; creatinine 0.11mg/dl; lactate 6.1 mmol/L, international normalized ration (INR) 3.52, activated partial thromboplastin time (aPTT) ratio 3.05, and antithrombin (AT) III 17%
- Peripheral venous access was obtained for the administration of fluids (NaCl 0.9 % with 10 ml/h), prothrombin complex (300 UI) and vitamin K (10 mg)
- Ten hours after admission, the patient developed tachypnea, anuria and her neurological status deteriorated severely
- Despite the initial therapies, laboratory data indicated a rapid worsening of coagulation parameters: INR and aPTT ratio were not measurable, PLT 68,000/mm3, Hb 7.6 g/dl, ATIII 7 %
- Simultaneously, she developed acute renal failure
- Following transfer to the Intensive Care Unit (ICU) with subsequent sedation and paralysis, the patient was intubated and mechanically ventilated in pressure control mode
- Additionally, immediate transfusion with red blood cells (RBC) was performed
- Chest radiography indicated pulmonary thickenings
- Blood gas analysis showed a pH of 7.44, $PaCO_2$ 23 mmHg, PaO_2 287 mmHg, lactate 2.4mmol/L, and base excess (BE) of -7.5 mmol/L

- Two peripheral intravenous catheters, one central venous catheter (left femoral vein), a right femoral arterial catheter for continuous pressure monitoring and a bi-lumen catheter for hemodialysis placed into the right femoral vein, were inserted. Of note, a significant amount of bleeding occurred when the catheters and a nasogastric tube were inserted, resulting in the immediate start of blood transfusions
- The thromboelastogram showed a value ("R") of 73 minutes, followed by administration of red blood cells 240 ml, fresh frozen plasma (FFP) 130 ml, platelets 100 ml, fibrinogen 150 mg, non-activated plasma derived factor VII (Provertinum, 400 UI) and human prothrombin complex (400 UI)
- After this initial (coagulation-focused) treatment, laboratory data showed a slight improvement in coagulation parameters: INR 4.27, PLT 13,000/mm3, fibrinogen 112 mg/dl, ATIII 13%, Hb 12.4 g/dl, Hct 34%, although aPTT was still not measurable
- Due to the development of acute renal failure, continuous renal replacement therapy (CRRT) was initiated
- With the rationale to reduce circulating bilirubin and ammonia plasma levels, a CytoSorb hemoadsorption cartridge was additionally integrated into the CRRT circuit

Treatment

- CytoSorb was used in combination with CRRT (Prismaflex System, Baxter, Germany) run in Continuous Veno-Venous Hemodiafiltration (CVVHDF) mode
- Applied CRRT dose: between 20-30 ml/kg/h
- Anticoagulation: regional citrate anticoagulation (RCA). Of note, after 3 hours of combined therapy, the decision was made to stop the RCA due to a rapid increase in lactate levels
- The priming procedure was performed with whole blood (100 ml)
- CytoSorb adsorber position: post-hemofilter

Measurements

- Bilirubin and urea plasma levels
- Pediatric Risk of Mortality score (PRISM III) and Sequential Organ Failure Assessment (SOFA) Score
- Overall clinical condition

Results

- After 18 hours of combined CRRT and CytoSorb hemoadsorption treatment, there was a massive reduction in bilirubin plasma concentrations (from 29.42 to 2.91 mg/dL) while urea levels decreased simultaneously
- Furthermore, there was a considerable improvement in the PRISM III (from 21 to 17) and SOFA Score (from 13 to 7)
- Importantly, during CVVHDF and CytoSorb treatment, the same dosage of catecholamines was administered without any worsening in the hemodynamics

Patient Follow-up

- After three days, the patient could be transferred to the national transplant center for liver transplantation which she received, and was discharged home after a further four months

Conclusion

- This is the first experience with the use of provertinum and CytoSorb in a low weight infant as a bridge to liver transplantation. Initiation of hemadsorption therapy was associated with a significant reduction in bilirubin and ammonia levels
- The authors report that this preliminary experience underlines the role of hemoadsorption in the management of severe hepatic failure in pediatric patients before liver transplantation
- Finally, the authors conclude with their opinion that, in general, CytoSorb application in neonates and pediatrics is very useful in many pathological conditions