Liver Transplantation in a Sickle Cell Disease Patient

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Introduction:

Sickle cell disease (SCD) is a recessive autosomal transmitted rare hematologic disease in which S-hemoglobin results from mutation 11th chromosome short arm. Patients develop chronic anemia and chronic pain crisis due to vascular occlusion in organs such as the liver. SCD affects an estimated 1 in 600 African American children, in Mexico is more prevalent in the Caribbean and Gulf coast. Hepatomegaly and biochemical abnormalities is usually common as a result of hemolysis and hepatocellular injury. Most patients have increased levels of unconjugated bilirubin, as well as aspartate aminotransferase (AST), alanine aminotransferase (ALT).

Sickle cell chronic hepatopathy may develop as a result of sickling process and multiple red blood cell transfusions which is believed secondary to iron overload.

There is a few publications of liver transplantation in SCD, the reported mortality rate arises 60% most of them in the first 6 months after liver transplantation. The major complication is graft thrombosis and infarction, pyogenic liver abscess, pulmonary embolism and multiorgan failure.

Methods:

We describe a 21 years old female patient with Child-Pugh C cirrhosis, with SCD diagnosis at age of 3. Multiple transfusion and iron chelants were given through childhood, at age of 7 an splenectomy was done and at age of 21 a cholecystectomy with liver biopsy revealed cirrhosis with large amounts of iron deposits. Acute on chronic liver failure with bilirubin of 40 mg/dl required liver dialysis (MARS), and then she was referred to the liver transplant department for study protocol.

A novel strategy of periodic whole blood exchange was indicated with the aim of maintain S-hemoglobin below 20%. After three months in the waiting list the patient received a liver transplantation from deceased donor with a whole blood exchange just before the procedure. At that time liver cirrhosis Child-Pugh C and Model of End Stage Liver Disease (MELD) of 21.

Results:

Liver transplantation was uneventful with full recovery and discharged with good liver function.

During the first 3 months after liver transplantation small areas of liver infarction were seen mainly in the right lobe, but without need of further intervention.


Conclusion:

The patient accomplished 32 months survival with normal liver function and no rejection episodes, she continues with periodic S-hemoglobin determination to maintain it below 20%.

Whole blood exchange is an effective strategy to reduce the risk of graft thrombosis in LT.