Decompensating Infantile Hemangioendothelioma: A Multidisciplinary Approach to Optimize a Three Month Patient for Liver Transplantation

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Background

Infantile hepatic hemangioendothelioma is the most common benign liver tumor in children. About 10 percent of these tumors require treatment. Treatment for this tumor ranges from liver transplant in extreme cases to TACE where extrahepatic disease is present. Medication like propranolol has also become important in the treatment to halt hemangioma proliferation. Our case examines the extreme range of this spectrum where a three month old girl has near complete replacement of her liver with a hepatic hemangioendothelioma requiring a multidisciplinary approach to be able to optimize her for liver transplant.

Material and Methods

The authors have no conflicts of interests or disclosures for this case report.

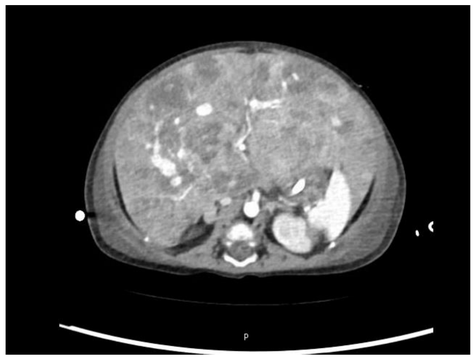

Fig. 1 Pre-operative CT scan illustrating markedly enlarged liver with diffuse heterogeneous densities, with prominent peripheral arterial enhancement, consistent with hemangioendothelioma.
Liver volume: 597 cm3

Case Report

A three month girl, former 38 week with IUGR, presented to Medstar Georgetown University Hospital (MGUH) for liver transplant due to infantile hepatic hemangioendothelioma. She was noted to have an enlarging abdomen at her 6-week wellness visit by her pediatrician. A CT scan at that time showed near complete replacement of the liver with a hepatic hemangioendothelioma and started on steroids at that time. Due to a lack of steroid response, she was started on a course of vincristine and sirolimus to shrink the mass. She was also started on propranolol one month prior to liver transplant due to its vasoconstrictive properties.

Prior to arrival to MGUH, she was noticed to have increasing respiratory effort and hypoxic episodes. She was intubated and had difficulties weaning from the ventilator. Her thyroid panel demonstrated a type 3 deiodinase and consumptive hypothyroidism. This massive hypothyroidism was secondary to a consumptive hypothyroidism by her tumor. At that time, her echocardiogram showed a hyperdynamic biventricular function with increase flow over the ascending aorta and pulmonary outflow tract as well a an enlarged left ventricle. As a result, her respiratory distress was believed to be cardiogenic in nature due to her hypothyroid state.

The patient was automatically placed on the transplant list as a status 1b, while a multidisciplinary approach was initiated to optimize the patient for liver transplantation. Since her admission, she was extubated after the tumor shrunk and thyroid function was normalized. Her repeat echocardiogram showed significant improvement with no heart failure or pulmonary hypertension.

A few weeks later she was given a left lateral segment orthotopic liver transplant via left hepatic artery to recipient left hepatic . Daily liver ultrasounds post-transplant for 5 days showed stable graft function

Case Report

with good flow. Propranolol and levothyroxine were eventually discontinued and the patient was successfully extubated on postoperative day two.

Results/Conclusion

This case report illustrates the importance of multidisciplinary approach in the optimization of complex patients for liver transplant. This patient would not have been able to optimized for liver transplant without the teamwork between the PICU, transplant surgery, anesthesiology, hepatology, cardiology, and endocrinology teams. Because of this collaborative effort, the patient was able to get her thyroid and heart function optimized while at the same time getting listed as a status 1b for liver transplant to be able to get her a liver for definitive treatment for her hemangioendothelioma.

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