

Hepatoblastoma

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Abstract

Hepatoblastoma is the most common primary hepatic malignancy of childhood. Patients most commonly present with abdominal distension and an elevated alpha-fetoprotein. Patients should be imaged via MRI using a hepatobiliary contrast agent. The PRE-Treatment EXtent of tumor staging systems is used to assess all pediatric liver tumors. Survival rates have improved over the past several decades. While tumor resection is required for a cure, patients also receive neoadjuvant chemotherapy.

Keywords: abdomen, neoplasm, liver

Case Summary

An infant presented to the emergency department with 10 days of abdominal distension, decreased oral intake, and one day of emesis. On laboratory evaluation, alpha-fetoprotein (AFP) was markedly elevated at 1,500,000 ng/mL (normal < 8 ng/mL).

Imaging Findings

Abdominal radiograph (Figure 1) showed an upper abdominal mass. This was confirmed to be a solid mass arising from the left lobe of the liver via US (Figure 2). Subsequent abdominal MRI (Figure 3) showed a large mass arising from the left lobe of the liver within the left medial and lateral sections.

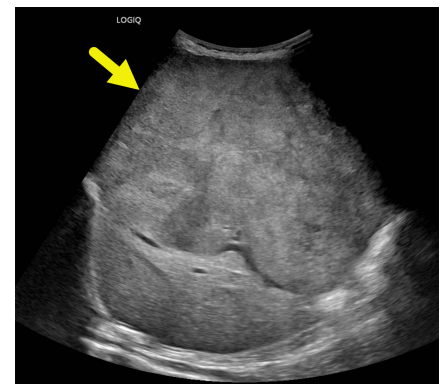
Diagnosis

Hepatoblastoma (HB)

Figure 1. Abdominal radiograph showing a mass within the right upper abdomen. The mass displaces loops of bowel inferiorly into the left.



Figure 2. Longitudinal image from abdominal US showing a large soft-tissue mass (arrow) arising from the left lobe of the liver.



Discussion

HB is the most common primary malignant hepatic tumor of childhood affecting 1 per 1.2 million children.¹ The tumor affects young children and is mostly diagnosed in the first 3 years of life.² HB is

more common in children born prematurely with a very low birth weight.³ It is also associated with familial adenomatous polyposis, trisomy 18, and the overgrowth syndromes (such as Beckwith-Wiedemann syndrome and Simpson-Golabi-Behmel syndrome).⁴

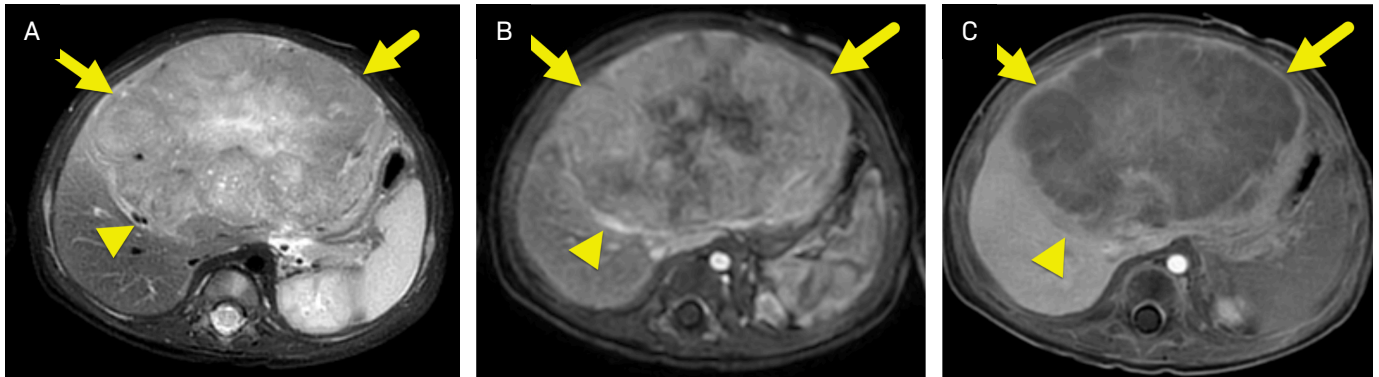
Children with HB most often present with abdominal fullness but may also have symptoms of vomiting, failure to thrive, or, rarely, jaundice. At laboratory evaluation, the AFP level is markedly elevated, often with values much higher than 100,000 ng/mL and in some patients >1,000,000 ng/mL.²

Abdominal US is the initial imaging modality performed in a child with an abdominal mass.¹ The US confirms the presence of the mass and helps identify the

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Disclosures: The authors have no conflicts of interest to disclose. None of the authors received outside funding for the production of this original manuscript and no part of this article has been previously published elsewhere.

Figure 3. (A) Axial T2-weighted MRI showing a large left-sided solid hepatic mass (arrow). The mass is margined by the middle hepatic vein (arrowhead), making it a PRE-Treatment EXTent of tumor II. (B) Axial T1-weighted postcontrast images in the portal venous and (C) hepatobiliary phase of imaging showing the mass (arrow) margined along its medial aspect by the middle hepatic vein (arrowhead).



organ of origin. On US, HB appears as a heterogeneous mass that is hypoechoic to the background liver. Small cystic spaces and echogenic calcification can be present.¹ Color Doppler interrogation is used to evaluate the hepatic vasculature. Specifically, it is used to identify the hepatic veins, inferior vena cava, and portal veins and determine their patency.⁵

Once a liver mass is identified on US, MRI with a hepatobiliary contrast agent is recommended as the next imaging modality.⁶ The hepatobiliary agent helps better identify tumoral margins, improve radiologist diagnostic confidence, and identify additional lesions.⁶ Multiphase postcontrast imaging is performed to better assess the hepatic vasculature and identify potential tumor thrombus. On MRI, the tumor is heterogeneous with mild hyperintense T2 signal and mild hypointense T1 signal.⁵ The enhancement pattern is variable and heterogeneous. However, the tumor typically enhances less than the background liver on all phases of enhancement.⁵ Abdominopelvic CT is not the imaging modality of choice. However, it can be performed in certain scenarios. When performed, imaging is recommended to be performed in the hepatic arterial and portal venous phases only. Noncontrast imaging and delayed phase imaging should not be performed.⁵ On CT, the tumor is heterogeneous and typically enhances less than the background liver. Tumoral calcification

may be present.⁵ Chest CT is recommended as lung metastases are present in 20% at diagnosis.⁵

The PRE-Treatment EXTent of tumor (PRETEXT) staging system is used to assess all pediatric liver tumors. This staging system is used to help risk-stratify patients prior to therapy. Additionally, it has been shown in multiple cancer cooperative group trials to help predict overall survival.⁷

The staging system is comprised of a PRETEXT group and annotation factors. The PRETEXT group is determined by calculating the number of contiguous liver sections that would have to be removed to completely excise the tumor.⁶ For example, a PRETEXT I tumor is one that is confined to either the right posterior or left lateral section. In this instance, only one section of the liver would need to be resected to completely excise the tumor. Details related to the anatomic boundaries and PRETEXT groups are included in the most recent PRETEXT manuscript.

Once the PRETEXT group is determined, the radiologist should identify any potential annotation factors. These factors are items that increase surgical complexity and include hepatic venous or inferior vena cava involvement, portal venous involvement, extrahepatic disease, multifocality, tumor rupture, caudate lobe involvement, lymph node metastasis, and distant metastasis.⁸ Each of these terms is defined in detail in the most recent PRETEXT manuscript.

HB cannot be cured without surgical resection. Surgical resection is usually combined with neoadjuvant chemotherapy drugs such as doxorubicin, cisplatin, vincristine, and 5-fluorouracil.⁹ Liver transplant is reserved for unresectable HB. These operations often have very successful outcomes.¹⁰ In standard-risk tumors, survival rates are >90% thanks to improved surgical techniques and chemotherapy.⁸

Conclusion

HB is the most common primary hepatic malignancy of childhood. Patients most commonly present with abdominal distension and an elevated AFP. Patients should be imaged via MRI using a hepatobiliary contrast agent. The PRETEXT staging system is used to assess all pediatric liver tumors. Survival rates have improved over the past several decades. While tumor resection is required for a cure, patients also receive neoadjuvant chemotherapy.

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