Radiology Case Reports 19 (2024) 5804–5808

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Abdominal pain and distension in a 4-years-old child revealing an hepatoblastoma ☆

*Kenza Berrada**, *Ibtissam El Ouali*, *Yahya El Harass*, *Lina Belkouchi*, *Nazik Allali*, *Latifa Chat*, *Siham El Haddad*

Department of Pediatric Radiology, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco

ARTICLE INFO

Article history:
Received 22 July 2024
Revised 30 July 2024
Accepted 31 July 2024

Keywords:
Abdominal mass
Abdominal pain
Pediatric
US
CT
MRI
PRETEXT

ABSTRACT

Hepatoblastoma (HBL) stands as the primary liver tumor most frequently encountered in children, typically identified within the initial 5 years of life. Cases involving patients older than 5 years are very rare. We report the case of a 4-year-old male child who presented to the emergency department with acute onset abdominal pain and fever. Clinical examination revealed significant abdominal distension, correlated with an abdominal mass later confirmed.

Introduction

Hepatoblastoma (HBL) stands as the most frequently encountered primary liver tumor in children. It is typically identified within the initial 5 years of life. Cases involving patients older than 5 years are very rare. We report the case of a 4-year-old male child who presented to the emergency department with acute onset abdominal pain and fever. Clinical examination revealed significant abdominal distension, correlated with an abdominal mass later confirmed.

Case report

We report the case of a 4-year-old who presented to the emergency department with a sudden onset of abdominal pain and fever.

The initial clinical examination showed a significant abdominal distension with a localized mass syndrome observed in the right flank.

An initial abdominal ultrasound revealed a large right hepatic mass with a smooth slope connecting to the hepatic parenchyma, grossly rounded, poorly defined, exhibiting

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

* Corresponding author.
E-mail address: knouz.berrada@gmail.com (K. Berrada).

https://doi.org/10.1016/j.radcr.2024.07.185

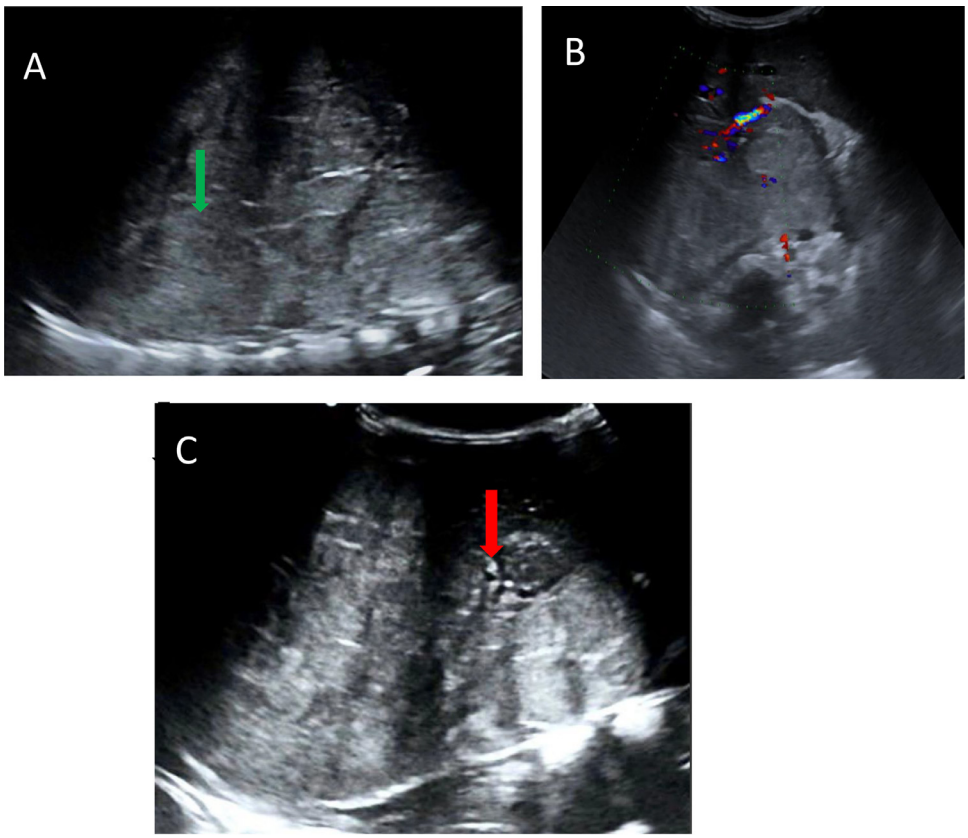

Fig. 1 – Abdominal ultrasound revealing a large right hepatic mass with a smooth slope connecting to the hepatic parenchyma, grossly rounded, poorly defined, exhibiting heterogeneous isoechoic echotexture (A: red arrow) containing areas of fluid and calcifications (C : red arrow), and taking moderately the color Doppler (B).

heterogeneous isoechoic echotexture containing areas of fluid and calcifications (Fig. 1).

Subsequent contrast-enhanced CT scan demonstrate a large hepatic mass, roughly rounded with polylobed contours, heterogeneous, containing calcifications and liquid areas, showing moderate enhancement after injection of contrast agent (Fig. 2). Axial CT section of lung window revealed a metastatic pulmonary nodule (Fig. 3).

For histological confirmation, an ultrasound-guided biopsy was performed, revealing an hepatoblastoma.

Discussion

Abdominal tumors rank as the third most common malignancies in children, following hematologic malignancies and central nervous system tumors. Among these, the 3 most prevalent abdominal solid tumors in children are neuroblastoma (NB), Wilms tumor (WT), and hepatoblastoma (HB), comprising approximately 5%, 4%, and 1% of all malignancies in children under 20 years of age. Due to their origin from embryonal or blastemal elements, these tumors are most frequently diagnosed in children under 5 years old, collectively accounting for around 14%, 10%, and 2% of malignancies within this age group [1].

Primary liver tumors constitute 15% of all abdominal tumors in childhood, with malignancy mean of 66% [2]. Hepatoblastoma is the most common [2], it presents a peak incidence between 2-5 years of age, and a prevalence slightly higher in boys than in girls [2]. Being infrequent in children older than 5 years, hepatoblastoma carries a worse prognosis due to late diagnosis, and hepatocellular carcinoma accounts for 87% of cases in this age range [2].

Although tumors are primarily sporadic, certain genetic syndromes are linked to a higher risk of HBL. These include Beckwith-Wiedemann Syndrome (BWS), familial adenomatous polyposis, and trisomy [3]. Screening recommendations for BWS typically entail abdominal ultrasound every 3 months from diagnosis until the age of 7 to 8 years, along with serum alpha-fetoprotein (AFP) measurement every 3 months until the age of 4 years [3].

Other factors contribute to the rising prevalence of HB. The improved survival of premature infants is linked to a higher incidence of HB, as very low birth weight is a known risk factor [1]. Other factors reported to correlate with a higher incidence of hepatoblastoma in children include preeclampsia, polyhydramnios or oligohydramnios, high maternal prepregnancy weight, and treatment for infertility in women. Also, some associations have been observed between tobacco smoking of both parents added to exposure to metals [2].

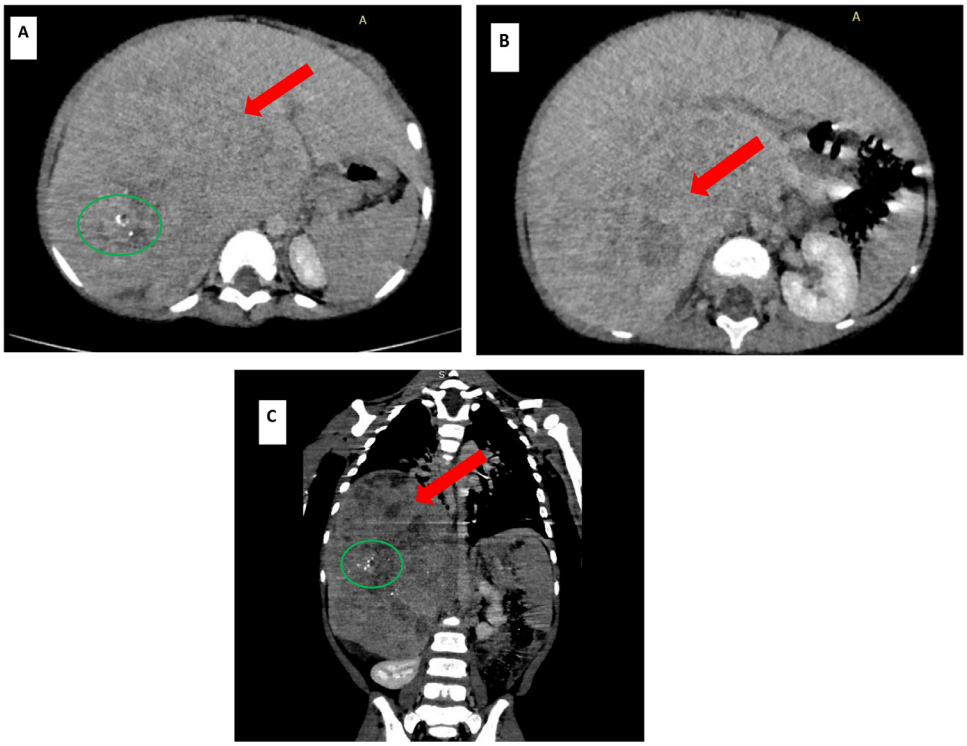

Fig. 2 – Axial (A and B) and coronal (C) enhanced CT scan demonstrate a large hepatic mass (red arrow), roughly rounded with polylobed contours, heterogeneous, containing calcifications (red circle) and liquid areas, showing moderate enhancement after injection of contrast agent.

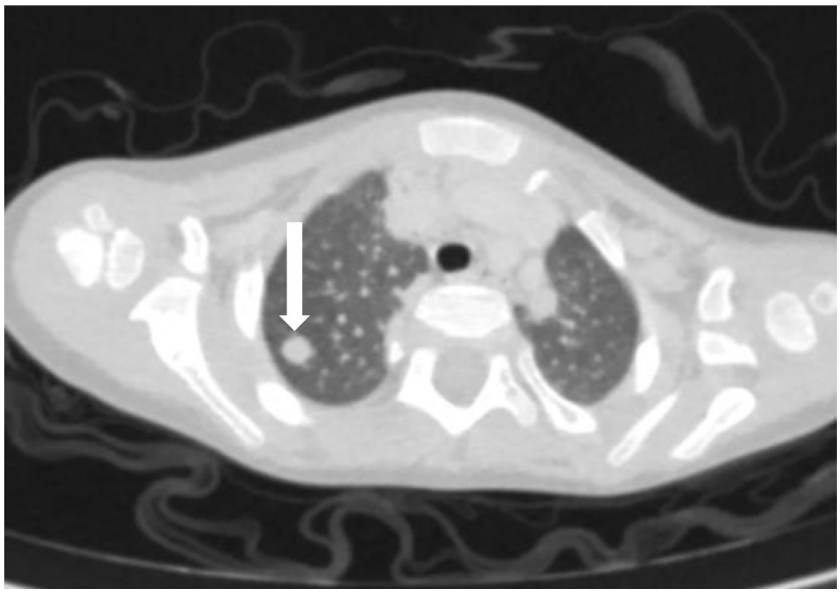

Fig. 3 – Axial CT section of lung window revealed a metastatic pulmonary nodule (white arrow).

Clinically, abdominal distension combined to a palpable mass are the most common symptoms. Sometimes abdominal discomfort, general fatigue, and loss of appetite or secondary anemia can be indicators. In cases of ruptured tumors, children typically exhibit vomiting, signs of peritoneal irritation, and severe anemia. Occasionally, rare instances may present with precocious puberty or virilization, attributed to β-human chorionic gonadotropin (HCG) secretion by the tumor [4].

Imaging is crucial for both the initial evaluation and the assessment of treatment response. Ultrasound (US) is often the first imaging modality performed, so many cases of hepato-

blastoma (HB) are initially identified using US. Primary tumors are typically large and exhibit variable echogenicity compared to the liver parenchyma. They may contain areas of necrosis, calcification, and hypoechoic fibrotic septa [5]. Once HB is suspected, further evaluation with MRI (or contrast-enhanced CT when appropriate) is recommended [6].

Due to its superior soft-tissue contrast, the availability of hepatobiliary contrast agents, and the capability to conduct dynamic contrast-enhanced imaging without ionizing radiation, MRI is the preferred cross-sectional imaging method for assessing HBL. It typically appears hypointense on T1-weighted and hyperintense on T2-weighted compared to normal liver parenchyma with restriction on DWI. On dynamic contrast-enhanced imaging, most HBL exhibit heterogeneous enhancement but to a lesser extent than normal liver tissue. A distinct demarcation usually exists between the tumor margin and hyperintense liver parenchyma. This demarcation aids in accurate anatomical tumor localization, assessment of the relationship between the tumor and vascular and biliary structures, and identification of additional hepatic disease foci [1].

CT can serve as a valuable imaging tool in assessing pediatric patients with known or suspected hepatoblastoma. Imaging the primary liver tumor using CT offers advantages over MRI, including superior spatial resolution, which allows for better delineation of small structures like hepatic vessels. Additionally, CT enables evaluation of the lungs during the same examination for the presence of metastatic disease, and it typically involves faster imaging with reduced sedation or anesthesia requirements [6].

On CT scans, hepatoblastoma (HB) typically appears well-defined and slightly hypoattenuating compared to the liver parenchyma. Calcifications are present in about 50% of tumors. Iodinated IV contrast material should also be used. Imaging of the upper abdomen, including the liver, should be performed in the late arterial phase, followed by imaging of the entire abdomen and pelvis in the portal venous phase. This dual-phase approach provides a comprehensive assessment of the liver's arterial and venous vasculature, which is essential for surgical planning. HB often shows heterogeneous enhancement, with tumor components exhibiting strong arterial enhancement that progresses to an overall enhancement slightly less than that of the liver parenchyma in the portal venous phase [1].

PRETEXT system was developed to standardize the imaging assessment and risk stratification of hepatoblastoma before the administration of neoadjuvant chemotherapy or tumor resection. Initially introduced in 1992 by the SIOP Epithelial Liver Tumor Study Group (SIOPEL), it has undergone several revisions over the years. The most recent version, published in 2018, represents the latest advancements in the field [6].

The PRETEXT group (I, II, III, or IV) is determined based on the number of contiguous liver sections that need to be resected to completely remove the tumor. Once the PRETEXT group is determined, annotation factors are applied to describe various aspects such as the tumor's impact on hepatic vascular structures, presence of extrahepatic disease contiguous with the main liver tumor, tumor multifocality, tumor rupture, involvement of the caudate lobe, and the presence of lymph node and distant metastases. Factors linked to poorer outcomes include advanced PRETEXT group, involvement of hepatic or portal venous structures, presence of extrahepatic disease, multifocality of the primary tumor, tumor rupture, and metastatic disease [7].

Histological confirmation of hepatoblastoma remains the examination of choice to confirm the diagnosis. HBL is an embryonal tumor originating from hepatic precursor cells, displaying morphological characteristics reflecting various cell types with different degrees of differentiation. It encompasses several tumor subtypes, predominantly epithelial in origin [8,9].

Serum alpha-fetoprotein (AFP) serves as the primary clinical marker for HBL. It plays a crucial role in monitoring treatment response, and relapse. However, certain variants of both HBL and hepatocellular carcinoma (HCC) may exhibit low or normal AFP levels. These variants, such as rhabdoid tumors, yolk sac tumors, mesenchymal hamartomas, infantile hemangioma endothelioma and focal nodular hyperplasia, may display distinct histological features and a poorer prognosis [10,11].

The differential diagnosis of malignant liver tumors in pediatric patients aged between 6 months and 6 years includes hepatocellular carcinoma (HCC), angiosarcoma and undifferentiated embryonal sarcoma of liver.

Hepatocellular carcinoma (HCC) is the second most common liver cancer in children. Its occurrence is most pronounced among adolescents, unlike hepatoblastoma, which primarily affects children under 5 years old. HCC can either be developed de novo on a nonpathological liver or on a cirrhosis liver. Calcifications are less common in HCC compared to other hepatic tumors. A large hypervascularized mass with a wash-out phenomenon are classic feature of HCC on MRI or CT, with 95% positive predictive value. The extent of the tumor, resectability, and the presence of vascular invasion and/or metastases are preferred to be assessed by CT [12].

Pediatric hepatic angiosarcoma is a rare malignant vascular tumor, comprising approximately 0.3% to 2.5% of liver tumors in children. It can manifest from the neonatal period up to adolescence with a female predominance, and reported median age of onset around 40 months [12]. Histological confirmation of hepatic angiosarcoma remains the examination of choice to assess the diagnosis. Imaging features are no specific; sometimes resembling to other vascular liver lesions [13].

Being considered as the third most common malignant hepatic tumor in childhood, undifferentiated embryonal sarcoma of the liver comes after hepatoblastoma and hepatocellular carcinoma [14]. This aggressive tumor of mesenchymal origin [15], classically presents in children from the age of 6 to 10 with no gender predilection. Due to the high water content of the prominent myxoid stroma, undifferentiated embryonal sarcoma appears as well-demarcated, predominantly hypoattenuated cystic mass with internal septations that slightly enhance after injection of the PDC, with multiple internal septa [16].

The cornerstone of hepatoblastoma (HB) treatment is surgical resection, whether performed upfront, after neoadjuvant chemotherapy, or through total hepatectomy followed by liver transplantation.

Currently, there are no standardized recommendations for imaging follow-up . Typically, patients with abnormal AFP levels post-treatment, including after surgery, are considered to have residual or recurrent disease. When levels of AFP return to normal after treatment, no additional imaging is required [1].

Conclusion

In conclusion, hepatoblastomas are the most common liver tumors found between 6 months and 6 years of age. Imaging plays an important role in diagnostic guidance, pre- and post-treatment assessment, and follow-up.

Patient consent

Written informed consent for publication was obtained from patient.

RÉFÉRENCES

[1] Morin CE, Artunduaga M, Schooler GR, Brennan RC, Khanna G. Imaging for staging of pediatric abdominal tumors: an update, from the AJR special series on cancer staging. Am J Roentgenol 2021;217(4):786–99.

[2] Pateva IB, Egler RA, Stearns DS. Hepatoblastoma in an 11-year-old: case report and a review of the literature. Medicine (Baltimore) 2017;96(2):e5858.

[3] an TY, Amor DJ. Tumour surveillance in Beckwith-Wiedemann syndrome and hemihyperplasia: a critical review of the evidence and suggested guidelines for local practice. J Paediatr Child Health 2006;42:486–90.

[4] Hiyama Eiso. Pediatric hepatoblastoma: diagnosis and treatment. Translational Pediatrics 2014;3(4):293.

[5] Chung EM, Lattin GE, Cube R, Lewis RB, Marichal-Hernández C, Shawhan R, et al. From the archives of the AFIP: pediatric liver masses: radiologic-pathologic correlation part 2. Malignant tumors. Radiographics 2011;31(2):483–507.

[6] Schooler GR, Squires JH, Alazraki A, Chavhan GB, Chernyak V, Davis JT, et al. Pediatric hepatoblastoma, hepatocellular carcinoma, and other hepatic neoplasms: consensus imaging recommendations from American College of Radiology Pediatric Liver Reporting and Data System (LI-RADS) Working Group. Radiology 2020;296(3):493–7.

[7] Towbin AJ, Meyers RL, Woodley H, et al. 2017 PRETEXT: radiologic staging system for primary hepatic malignancies of childhood revised for the Paediatric Hepatic International Tumour Trial (PHITT). Pediatr Radiol 2018;48(4):536–54.

[8] Ng K, Mogul DB. Pediatric liver tumors. Clin Liver Dis 2018;22(4):753–72.

[9] Ranganathan S, Lopez-Terrada D, Alaggio R. Hépatoblastome et carcinome hépatocellulaire pédiatrique : une mise à jour. Pathologie pédiatrique et développementale 2020;23(2):79–95.

[10] De Ioris M, Brugieres L, Zimmermann A, Keeling J, Brock P, Maibach R, et al. Hepatoblastoma with a low serum alpha-fetoprotein level at diagnosis: the SIOPEL group experience. Eur J Cancer 2008;44(4):545–50.

[11] Trobaugh-Lotrario AD, Tomlinson GE, Finegold MJ, Gore L, Feusner JH. Small cell undifferentiated variant of hepatoblastoma: adverse clinical and molecular features similar to rhabdoid tumors. Pediatr Blood Cancer 2009;52(3):328–34.

[12] Lucas B, Ravishankar S, Pateva I. Pediatric primary hepatic tumors: diagnostic considerations. Diagnostics 2021;11(2):333.

[13] Geramizadeh B, Kashkooe A, Bahador A, Dehghani SM, Shamsaeefar A, Kazemi K, et al. Pediatric hepatocellular carcinoma, a single center study from the South of Iran: case series. Hepatitis Monthly 2017;17(8):15.

[14] Martins-Filho SN, Putra J. Hepatic mesenchymal hamartoma and undifferentiated embryonal sarcoma of the liver: a pathologic review. Hepatic Oncol 2020;7(2):HEP19.

[15] Stocker JT, Ishak KG. Undifferentiated (embryonal) sarcoma of the liver. Report of 31 cases. Cancer 1978;42(1):336–48.

[16] Putra J, Ornvold K. Undifferentiated embryonal sarcoma of the liver: a concise review. Arch Pathol Lab Med 2015;139(2):269–73.