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A RARE MALIGNANT HEPATIC TUMOR OF CHILDHOOD: TRANSITIONAL LIVER CELL TUMOR REVISITED

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Transitional liver cell tumor is an extremely rare entity and has a poor prognosis. It has similar histopathologic findings with hepatoblastoma and hepatocellular carcinoma. Up to now, only 10 cases have been reported in the literature. We report on an 8-year-old boy with histologically proven transitional liver cell tumor and describe the pertinent radiological findings.

Key-word: Liver neoplasms, in infants and children.

Two-thirds of primary liver tumors in the pediatric population are malignant, and malignant primary hepatic tumors account for 1%-2% of all childhood cancers (1). Primary malignant liver tumors in children comprise a heterogeneous group of neoplasms, the majority of them being hepatoblastomas (HBL) and hepatocellular carcinomas (HCC). HBL occurs almost exclusively in patients younger than 5 years old, whereas HCC develops in older children and adolescents (2). On the other hand, transitional liver cell tumor (TLCT) is an aggressive tumor, which is histologically neither typical HBL nor HCC; however, it has several pathological findings in common with those tumors. To the best of our knowledge, there are only 10 pediatric patients reported with TLCT in the literature (3). In this regard, we report an 8-year-old boy with histologically proven TLCT and describe the pertinent radiological findings.

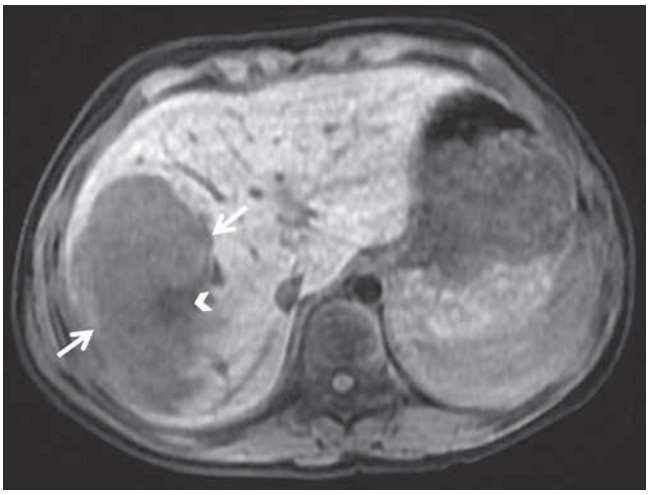

Fig. 1. — Axial T1-weighted image shows hypointense tumor with lobulated margin (arrows) and cystic-necrotic area in the central part of the tumor (arrowhead).

Case report

An 8-year-old boy presented with a one-month history of right upper abdominal pain. He described a fall episode one-month before the onset of abdominal pain. The medical history was otherwise unremarkable.

Physical examination revealed mild hepatomegaly. An abdominal ultrasound (US) showed mild hepatomegaly and heterogeneous mass with solid and cystic components in the right lobe of the liver (10 cm in anterior-posterior and 8 cm in transverse diameter). The right portal vein was encased within the mass and there was also 2 × 1 cm portal lymphadenopathy. Laboratory tests showed anemia, leukocytosis (with neutrophilia) and elevated erythrocyte sedimentation rate. Liver function tests were normal. Serum α-fetoprotein (AFP) level was significantly elevated 37241.67 IU/mL (N: 0-6.67). Magnetic resonance imaging (MRI) revealed a multilobulated-large (9.5 cm in craniocaudal, 10.5 cm in anterior-posterior and 7 cm in transverse diameter), heterogeneous solid mass in the right liver lobe. The tumor involves segments V, VII and VIII (PRETEXT Classification II) (4). The tumor exhibits central necrotic areas with low signal intensity on T1-weighted and high signal intensity on T2-weighted images (Fig. 1 and 2). The tumor enhanced slightly, but less than adjacent liver parenchyma after gadolinium administration (Fig. 3) and showed diffusion restriction on diffusion-weighted images. Encasement of the right hepatic vein could also be seen.

The US guided percutaneous biopsy was performed and histological examination revealed a tumor composed of cellular components with features of both HBL and HCC (Fig. 4). The tumor was diagnosed as TLCT. The patient received chemotherapy. On repeat abdominal MRI (3 months later), the size of the tumor was found not to have changed -with further elevated AFP levels (93095.40 IU/ml). Thereafter, a right extended hepatectomy was performed. One week after surgery, AFP level dropped to 11901.30 IU/ml. The patient also received postoperative chemotherapy. Two months after the operation, abdominal US showed a 1 cm hypoechoic mass in the left hepatic lobe parenchyma. There was a 2×2 cm solid mass on the anterior aspect of the left hepatic lobe outside the liver capsule and a 4 × 3.5 cm solid mass in the right lower quadrant near the incision scar in the subcutaneous tissue. Doppler US revealed increased vascularity in both

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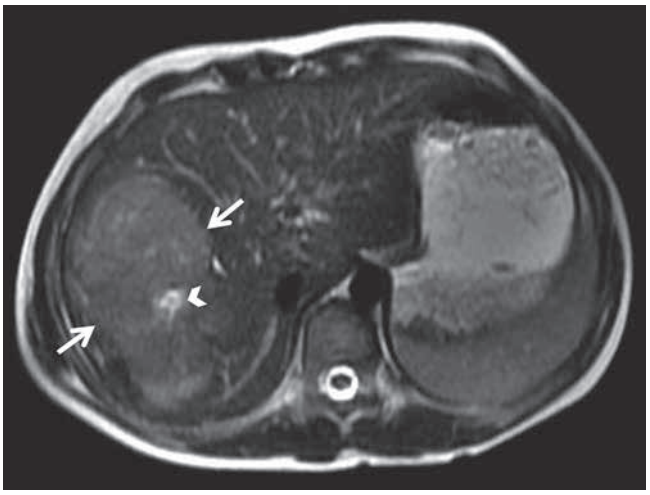
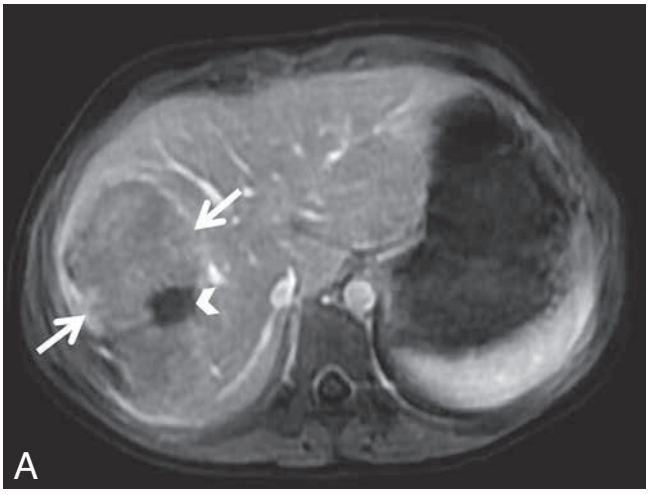


Fig. 2. — Axial T2-weighted MR image demonstrates a large hyperintense tumor in the right liver lobe (arrows). Note also the cystic-necrotic area in the central part of the tumor (arrowhead).



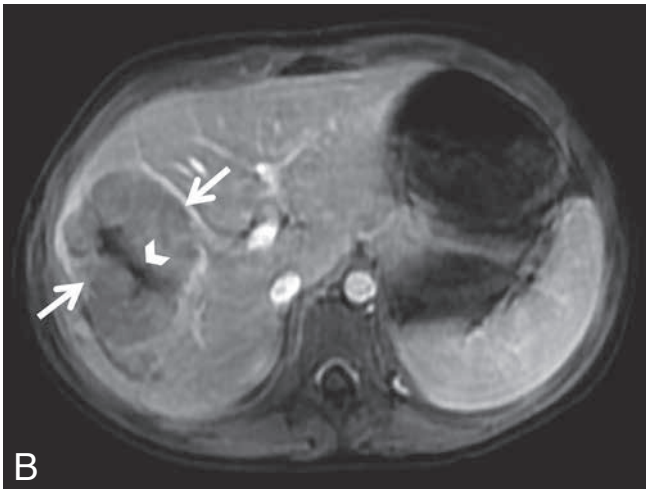


Fig. 3. — Axial gadolinium-enhanced T1-weighted MR images show heterogeneous enhancement of the tumor which is hypointense relative to the normal liver parenchyma (arrows in A and B). There is lack of enhancement of the central necrotic area (arrowheads in A and B).

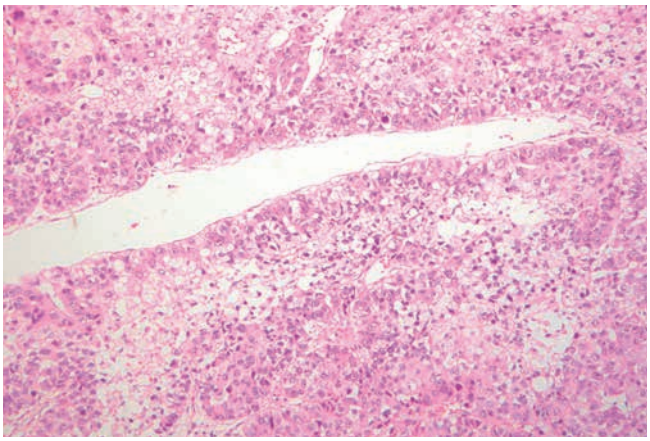


Fig. 4. — Hepatocellular carcinoma component with trabecular pattern composed of cells with high nucleus-cytoplasmic ratio (Hematoxylin and Eosin. Original magnification: ×100).

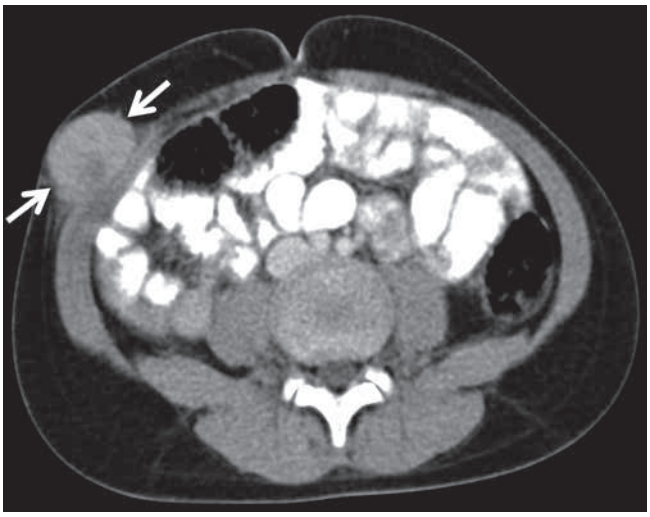


Fig. 5. — Two months after operation post contrast axial CT scan demonstrates a solid lesion in the right abdominal lateral wall (arrows).

lesions. Thorax and abdominal contrast enhanced CT showed multiple pulmonary metastases and multiple hypodense lesions in the left liver lobe, a solid mass in the anterior abdominal wall within the rectus abdominis muscle and another solid lesion in the right abdominal lateral wall (Fig. 5). At the last follow-up, AFP level was 17968.65 IU/ml.

Discussion

Primary malignant liver cell tumors in children represent a heterogeneous group, whereby HBL is seen predominantly in children less than 5 years of age, and HCC in older children and adolescents. Additionally, Prokurat and Zimmermann denoted a distinct group of malignant hepatocellular tumor named as TLCT, which

share common histological features with both of the aforementioned two types (5). They occur in older children and adolescents and markedly express beta-catenin, typically with a mixed nuclear and cytoplasmic pattern (5).

The usual presentation is that of a large or multifocal and/or unresectable primary mass most commonly seen in the right liver lobe. TCLT displays an expanding growth pattern, sometimes exhibiting a large central necrosis. In cases of TLCT, in contrast to HBL and HCC, the tumor displays a poor response to initial chemotherapy with higher serum AFP levels at diagnosis. In the previous reports, due to the giant size of the tumors, surgery was connected with many technical difficulties and resulted in a high percentage of patients in whom microscopic radicality of surgery was impossible (3).

Concerning the differential diagnosis, HCC usually occurs in children aged 10-14 years and 40-60% of the cases have elevated serum AFP levels (6). On post-contrast MR images, HCC typically demonstrates early arterial phase enhancement and may wash out with relative low signal intensity during the portal venous phase. If present, the tumor capsule is usually hypointense on T1- and T2-weighted images with delayed enhancement.

The imaging characteristics of HBL reflect its histologic composition. Histologically, HBL is classified into two types: the epithelial type and the mixed epithelial and mesenchymal type. Epithelial HBLs demonstrate a more homogeneous appearance, while mixed tumors are more heterogeneous in attenuation. At MR imaging, epithelial HBLs are homogeneously slightly hypointense on T1-weighted images and hyperintense on T2-weighted images relative to adjacent liver parenchyma. Mixed tumors demonstrate more heterogeneous signal intensity characteristics.

In our patient, the giant tumor included a central necrosis and serum AFP level was very high expected than HCC or HBL. MR images demonstrated slightly contrast enhancement after gadolinium administration but less than adjacent liver parenchyma. Diffusion restriction was seen on diffusion-weighted images.

In conclusion, presenting this rare case of ours, we underscore the importance of multidisciplinary approach for prompt diagnosis of TLCT whereby the onward treatment will be tailored accordingly.

References

1. Chung E.M., Lattin G.E. Jr., Cube R., et al.: From the archives of the AFIP: Pediatric liver masses: radiologic-pathologic correlation. Part 2. Malignant tumors. *RadioGraphics*, 2011, 31: 483-507.
2. Isaacs H. Jr.: Fetal and neonatal hepatic tumours. *J Pediatr Surg*, 2007, 42: 1797-1803.
3. Prokurat A., Chrupek M., Kaźmirczuk R., et al.: Primary malignant liver cell tumours in children different treatment strategies. *Ann Diagn Pediatr Pathol*, 2006, 10: 17-22.
4. Roebuck D.J., Aronson D., Clapuyt P., et al. International Childrhood Liver Tumor Strategy Group.2005 PRETEXT: a revised staging system for primary malignant liver tumours of childhood developed by the SIOPEL group. *Pediatr Radiol*. 2007, 37: 123-132.
5. Prokurat A., Kluge P., Kościesza A., Danuta P., Kappeler A., Zimmermann A.: Transitional liver cell tumors (TLCT) in older children and adolescents: A novel group of aggressive hepatic tumors expressing beta-catenin. *Med Pediatr Oncol*, 2002, 39: 510-518.
6. Moore S.W., Hesseling P.B., Wessels G., Schneider J.W.: Hepatocellular carcinoma in children. *Pediatr Surg Int*, 1997, 12: 266-270.