



PERGAMON

Short Communication

## Mesenchymal hamartoma of the liver

O. Konez\*, M. Goyal, P.K. Vyas, S.B. Boinapally

Department of Radiology, Children's Hospital Medical Center of Akron, Northeastern Ohio Universities College of Medicine (NEOUCOM), 300 Longwood Avenue, Boston, MA 02115, USA

Received 16 December 1999; accepted 27 June 2000

### Abstract

We present a multiseptated mesenchymal hamartoma of the liver in a 10-year-old male patient, a rare benign tumor of childhood. The characteristic ultrasound and CT appearances of this unusual tumor are reviewed. A single septal calcification associated with this tumor was demonstrated, an association which has not previously been reported. The differential diagnosis for cystic liver lesions is discussed in detail. © 2000 Elsevier Science Ltd. All rights reserved.

**Keywords:** Liver; Hamartoma; Mesenchymal; Calcification; CT; US

### 1. Case report

A 10-year-old African-American male presented to his pediatrician's office for a sports physical. He was found to have a right upper quadrant mass on physical examination and was admitted for further evaluation. His past medical and surgical history, family history, and review of systems were non-contributory. On physical examination, he had a non-pulsatile right upper quadrant mass extending 5 cm. below the costal margin. There was no apparent bruit. The remainder of the physical examination was unremarkable except for an eczematous rash on the anterior neck. All laboratory values, including alpha-fetoprotein (AFP) ( $<2.5$ ), were within normal range. The initial abdominal radiograph showed a large soft tissue mass within the right upper quadrant. A CT scan of the abdomen and pelvis revealed a large, multiloculated cystic liver mass with a single septal calcification (Fig. 1). An ultrasound examination of the upper abdomen was also performed for further characterization of this cystic mass, which showed low-level homogeneous echoes within a multiloculated liver mass (Fig. 2). The septal calcification, however, was not evident on the ultrasound images. By its characteristic sonographic appearance and clinical presentation, the diagnosis of mesenchymal hamartoma was made.

He was taken to the operating room, where he underwent exploratory laparotomy, partial resection of the right lobe of

the liver, core biopsies of the left lobe of the liver, cholecystectomy, and cholangiography. Frozen sections were done at the time of surgery. The operation as well as the postoperative care was without complications, and he was discharged home on postoperative day seven. Final pathology revealed that the tumor was a mesenchymal hamartoma of the liver. The initial prospective pathological examination of the mass did not identify any calcification. However, a retrospective review of the material showed a single focus of calcification within the mass corresponding to the calcification on CT. The core liver biopsies from the left lobe were normal.

### 2. Discussion

Mesenchymal hamartomas account for 22% of all benign liver tumors of childhood and they generally present in patients less than 2 years of age. The most common presentation is an asymptomatic abdominal mass [1,2]. Although abdominal radiographs usually demonstrate a non-specific large soft tissue mass in the right upper quadrant, this benign tumor can be diagnosed easily by its typical appearance of a multiseptated cystic mass on either sonographic examination or CT [2–6]. Predominantly solid appearing hepatic hamartomas have been, however, described in the literature [7]. By plain radiography alone, the foremost possibilities would include the more commonly seen entities such as metastatic liver disease from neuroblastoma, the

\* Corresponding author. Tel.: +1-617-355-6541; fax: +1-617-738-8216.

E-mail address: orhankonez@hotmail.com (O. Konez).



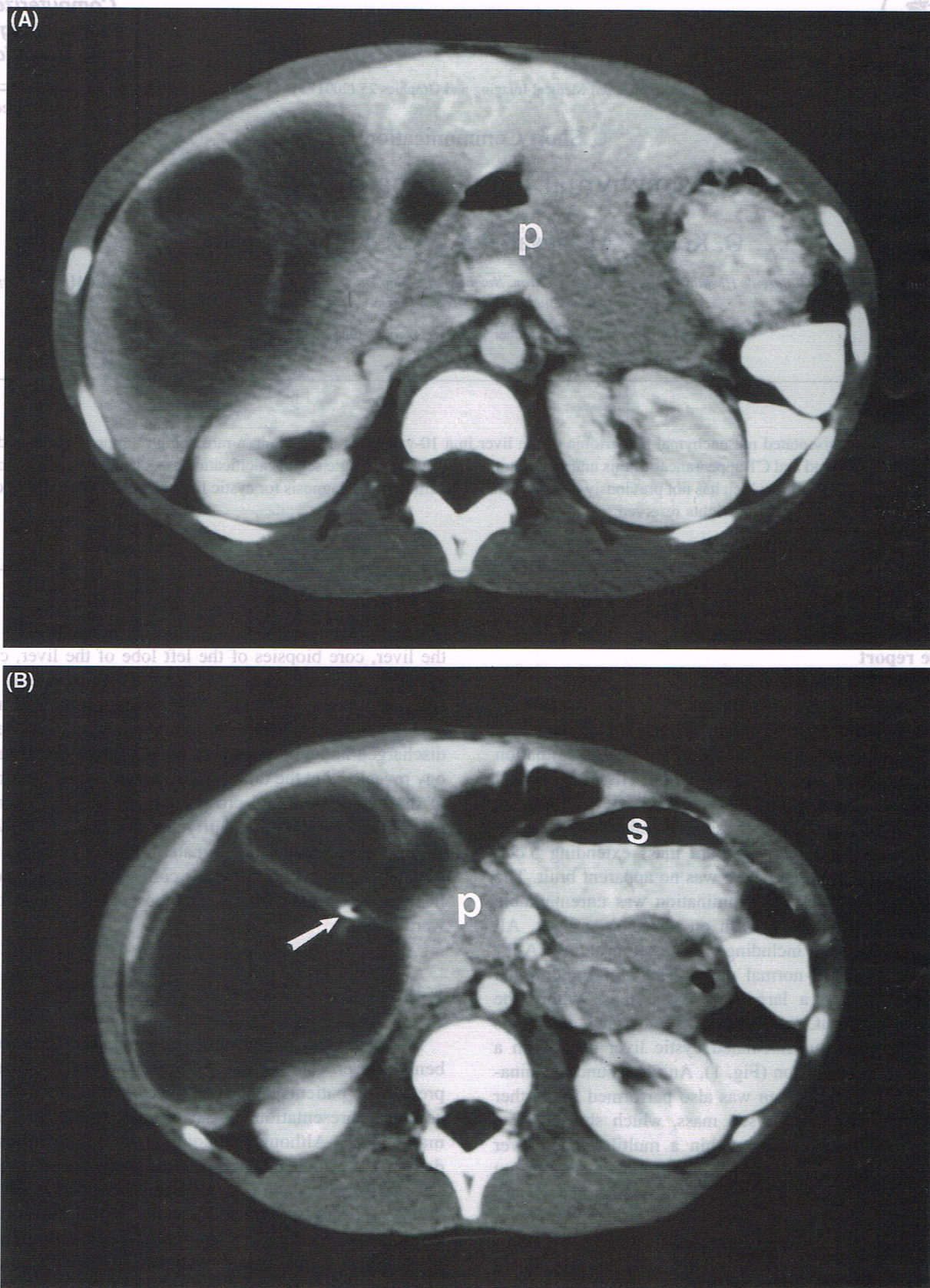


Fig. 1. (A, B) Two axial post-contrast (IV and oral) CT images through the upper abdomen demonstrating a well defined, lobulated and multiseptated cystic tumor mass in the right lobe of the liver. Note that the tumor mass is associated with a single rounded septal calcification (B)(arrow), which had a CT density of 262 HU. This association has not been reported previously. Midline structures are mildly displaced to the left. (p: pancreas, s:stomach).



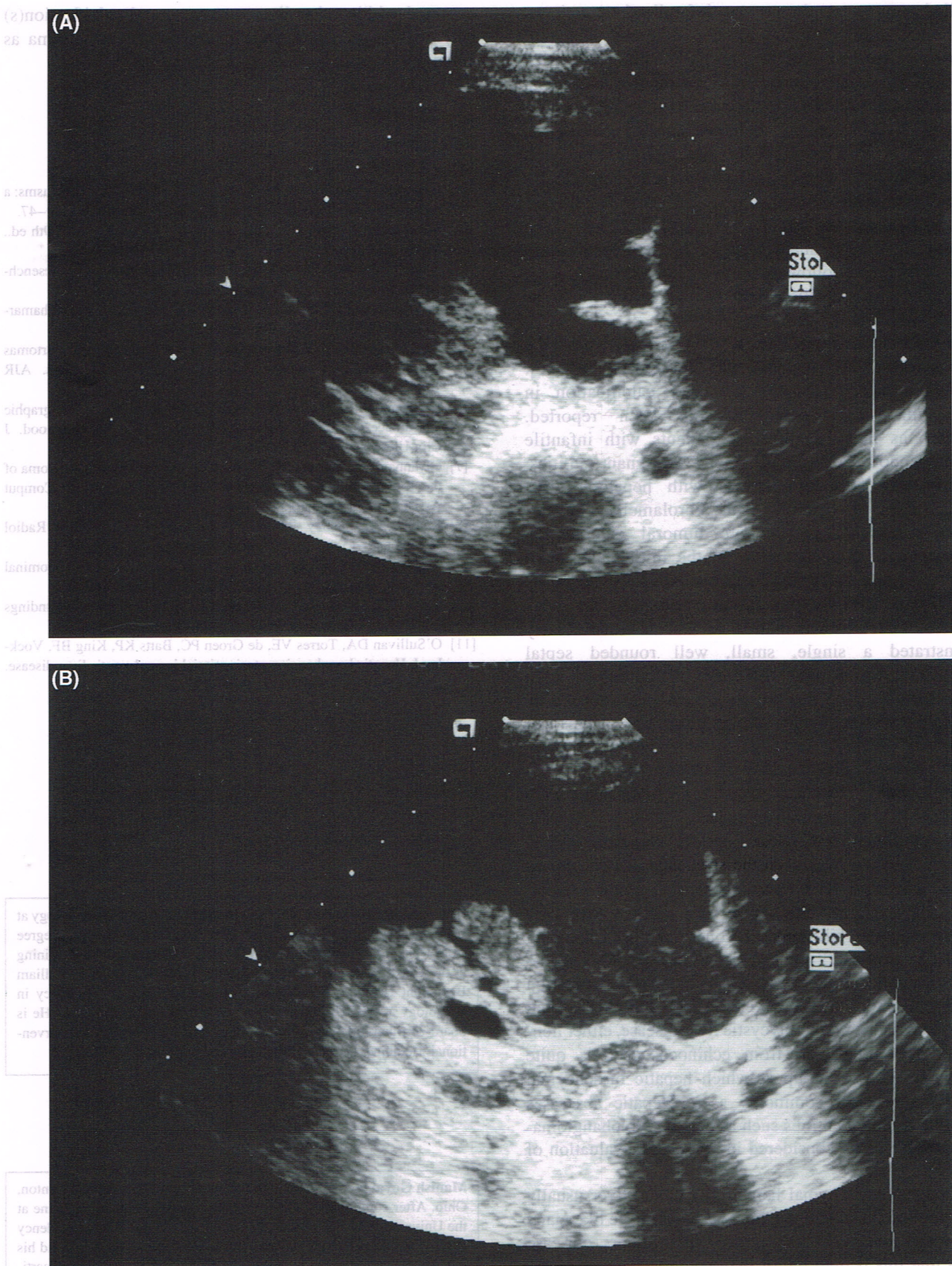


Fig. 2. (A, B) Two gray-scale transverse sonographic images of the right upper abdomen demonstrating multiseptated cystic mass in the liver with a low-level internal echogenicity, which is the typical appearance of hepatic mesenchymal hamartoma.



so-called pepper syndrome, or infantile hemangioendothelioma, although these entities, like mesenchymal hamartoma, are also rare beyond early childhood.

Mesenchymal hamartomas present most frequently as a single dominant cyst, occasionally some echogenic material within the cyst, secondary to blood, as well as, internal septae may be seen [2–5]. A less common appearance is that of a solid mass demonstrating vascular findings similar to those seen with other mesenchymally derived masses such as hemangioma or hemangioendothelioma [2,7]. CT typically shows enhancement of the non-cystic components of the mass following intravenous contrast infusion. These enhancing septations can be of varying thickness [2,5].

The presence of round hyperechoic nodules within the cystic spaces of a hamartoma was reported recently [6]. To our knowledge, septal (tumoral) calcification in mesenchymal hamartoma has not been reported. However, as many as 40% of patients with infantile hemangioendothelioma; and among malignant entities, approximately 50% of patients with hepatoblastoma and 35–40% of patients with fibrolamellar hepatocellular carcinoma demonstrate tumoral calcification [2]. No calcification was seen on the initial abdominal radiograph in our case, which was most likely due to its small size or perhaps due to obscuration by an overlying rib. The CT examination of the abdomen clearly demonstrated a single, small, well rounded septal calcification, which could be easily differentiated from other tumoral calcifications by its benign appearance. This calcification was not evident on the ultrasound images and was perhaps overlooked during real-time scanning. Hepatoblastoma typically demonstrates extensive amorphous calcification, while scattered punctate calcifications are commonly seen in neuroblastoma [2,8,9].

In the differential diagnosis, other cystic liver lesions such as polycystic liver disease or Caroli's disease should also be considered. Although the size, shape, and distribution of the cystic sacculi of Caroli's disease may vary greatly, calcium bilirubinate stones and/or enhancing vessels ("central dot") in these sacculi are strongly suggestive of this congenital disease entity [10]. The calcification seen in our case, however, is clearly not intraluminal, but rather septal in location. Polycystic liver disease, either non-parasitic (hereditary) or from echinococcosis, is quite unlikely in the age group at which hepatic mesenchymal hamartoma is most commonly seen. Hepatic abscesses and rare congenital entities such as hepatic lymphangiomas should also be considered in diagnostic evaluation of these cases [11].

Although mesenchymal hamartoma of the liver usually present within the first few years of life like many other pediatric benign and malignant liver tumors, our case proves that this benign tumor may come to medical attention, upon routine physical examination, as late as ten years of age. Although tumoral calcification is frequently

expected within other liver tumors, septal calcification(s) may also be seen in hepatic mesenchymal hamartoma as noted in our case.

## References

- [1] Helmberger TK, Ros PR, Mergo PJ, et al. Pediatric liver neoplasms: a radiologic–pathologic correlation. *Eur Radiol* 1999;9(7):1339–47.
- [2] Silverman FN, Kuhn JP. *Caffey's pediatric X-ray diagnosis*. 9th ed.. Mosby, 1993 (p. 954–67).
- [3] Rosenbaum DM, Mindell HJ. Ultrasonographic findings in mesenchymal hamartoma of the liver. *Radiology* 1981;138(2):425–7.
- [4] Federici S, Galli G, Sciutti R, Cuoghi D. Cystic mesenchymal hamartoma of the liver. *Pediatr Radiol* 1992;22(4):307–8.
- [5] Stanley P, Hall TR, Woolley MM, et al. Mesenchymal hamartomas of the liver in childhood: sonographic and CT findings. *AJR* 1986;147(5):1035–9.
- [6] Koumanidou C, Vakaki M, Papadaki M, et al. New sonographic appearance of hepatic mesenchymal hamartoma in childhood. *J Clin Ultrasound* 1999;27(3):164–7.
- [7] Giyannani VL, Meyers PC, Wolfson JJ. Mesenchymal hamartoma of the liver: computed tomography and ultrasonography. *J Comput Assist Tomogr* 1986;10(1):51–54.
- [8] Kenney PJ, Stanley RJ. Calcified adrenal masses. *Urol Radiol* 1987;9(1):9–15.
- [9] Araki T, Itai Y, Iio M. CT features of calcification in abdominal neuroblastoma. *J Comput Assist Tomogr* 1982;6(4):789–91.
- [10] Miller WJ, Sechtin AG, Campbell WL, Pieters PC. Imaging findings in Caroli's disease. *AJR* 1995;165:333–7.
- [11] O'Sullivan DA, Torres VE, de Groen PC, Batts KP, King BF, Vockley J. Hepatic lymphangiomas mimicking polycystic liver disease. *Mayo Clin Proc* 1998;73(12):1188–92.

**Orhan Konez** is a Fellow in the Division of Interventional Radiology at Children's Hospital, Boston, MA. After receiving his medical degree from Uludag University in Turkey, he had research fellowship training at the University of Washington, Seattle, WA and another at William Beaumont Hospital, Royal Oak, MI. He completed his residency in Radiology at NEOUCOM Residency Program in Canton, Ohio. He is board-certified in Diagnostic Radiology. His main interest is Interventional Medicine, Pediatric Radiology and MRI.

**Manish Goyal** is a body imaging Fellow at Aultman Hospital, Canton, Ohio. After completing a Research Fellowship in Nuclear medicine at the University of Pennsylvania, he completed his Radiology Residency at NEOUCOM residency program in Canton, Ohio in 1999. He did his medical schooling at the University of Delhi, India. He is board-certified in Diagnostic Radiology. His main interest is in Ultrasound and Mammography.



**Pranav K. Vyas** is Assistant Professor of Radiology at Northeastern Ohio Universities School of Medicine and Associate Radiologist at Children's Hospital Medical Center of Akron, in Akron. He obtained his undergraduate degree from the University of Pennsylvania and his medical degree from the University of Pittsburgh. Following residency in Diagnostic Radiology at Wayne State University, he completed a two-year fellowship in Pediatric Radiology and Pediatric Nuclear Medicine at Children's National Medical Center in Washington, DC. He is board-certified in Diagnostic Radiology, with Special Competence in Nuclear Radiology and a Certificate of Added Qualification in Pediatric Radiology. He was previously attending at Johns Hopkins Hospital and Instructor at the Johns Hopkins University School of Medicine. His clinical and research interests primarily include pediatric nuclear medicine and pediatric body MR and CT.

**Sujji B. Boinapally** is a third-year Resident at NEOUCOM Radiology Residency Program in Canton, Ohio. She graduated from Gandhi Medical College, India and worked as a post doctoral Research Fellow at Alton Ochsner Medical Foundation, New Orleans, Louisiana. She had an internship year at Lemuel Shattuck Hospital before she joined the Radiology Residency Program in Canton, Ohio.