ACG CASE REPORTS JOURNAL

IMAGE | PEDIATRICS

A Rare Case of Childhood Undifferentiated Embryonal Sarcoma of the Liver Managed Successfully

Gangadhar Rao Gondu, DNB[1], Vimalakar Reddy Eppa, DNB[2], Avinash Sowdepalli, DNB[1], Mohan Narasimhan, DNB, FACS[2], and Ramesh Ardhanari, MCh, FRCS[2]

[1]Department of General Surgery, Meenakshi Mission Hospital and Research Centre, Madurai, Tamil Nadu, India
[2]Department of Surgical Gastroenterology, Meenakshi Mission Hospital and Research Centre, Madurai, Tamil Nadu, India

Case Report

A 13-year-old boy presented with low-grade fever, upper abdominal pain, loss of appetite, and weight loss for 2 months, with a history of swelling in his upper abdomen for 15 days. On examination, a 17 x 20-cm intraabdominal, smooth, firm, non-tender swelling was noted occupying epigastric and left hypochondrium. The mass moved with respiration, and finger insinuation between mass and right coastal margin was not possible. There was no ascites and further examination was unremarkable. Liver function tests and α-fetoprotein were normal.

CT scan of the abdomen showed a homogenously enhancing mass lesion of 14.9 x 12.9 cm in the left lobe of the liver with multiple vascular structures traversing the lesion consistent with hepatocellular carcinoma (Figure 1). Non-anatomical left hepatectomy was done. Intraoperative examination revealed a 15 x 20-cm lesion arising from the inferior aspect of the left lateral segment of the liver, adhered to the stomach along the lesser curvature (Figure 2). Cut specimen showed solid tumor with cystic areas of hemorrhage and multiple vascular structures traversing the tumor (Figure 3).

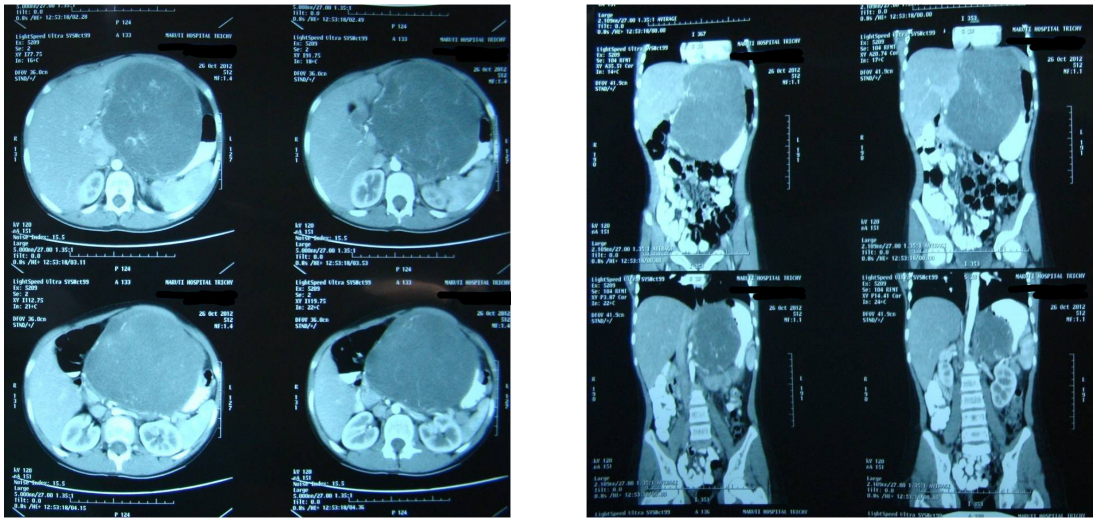

Figure 1. CT scan of upper abdomen (axial and coronal sections) showing homogenously enhanced mass lesion of 14.9 x 12.9 cm in left lobe of liver, with multiple vascular structures traversing the lesion suggestive of hepatocellular carcinoma.

ACG Case Rep J 2014;1(2):74–75. doi:10.14309/crj.2014.5. Published online: January 10, 2014.

Correspondence: Dr. Gangadhar Rao Gondu, DNB Resident, General Surgery, Meenakshi Mission Hospital And Research Centre, Lake Area, Melur Road, Madurai 625107 (drgangadhargondu85@gmail.com).

Postoperatively, 1 unit of packed red blood cell transfusion was given and the patient was discharged on postoperative day 8. Histopathological examination reported a tumor predominantly showing spindle cells arranged in sheets embedded in myxoid stroma. Vimentin was strongly positive. Desmin, carcinoembryonic antigen, and α-fetoprotein were negative. This suggested undifferntiated embryonal sarcoma of liver (USEL; Figure 4), a rare, highly malignant neoplasm that is more common in children. Bone scan was normal. Adjuvant chemotherapy was given. The patient is now asymptomatic on regular follow-up. We stress the role of histopathology and IHC in diagnosing UESL and of timely *en bloc* resection and postoperative chemotherapy in improving survival rates.

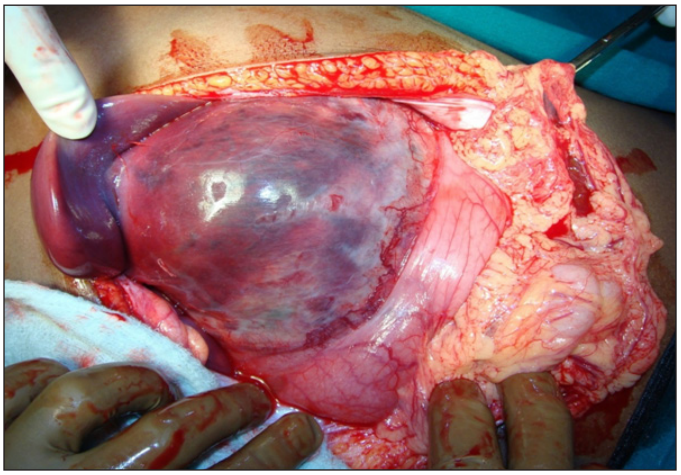

Figure 2. Intraoperative picture showing tumor arising from inferior surface of left lobe of liver, adhered to stomach along lesser curvature.

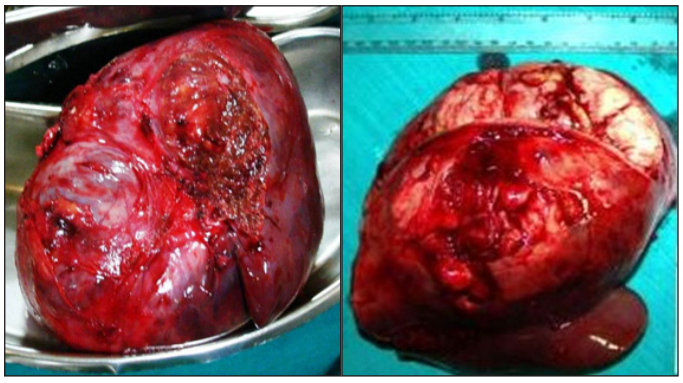

Figure 3. Excised specimen surrounded by capsule. Cut section showing myxoid stroma with areas of hemorrhagic necrosis.

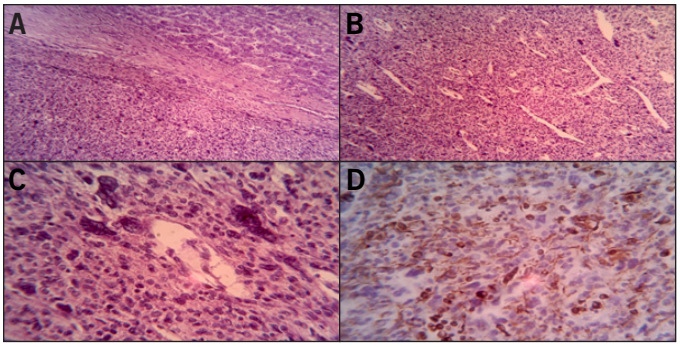

Figure 4. (A) 100x picture showing normal liver in right upper part and tumor in the lower part. (B) 100x tumor showing mixture of highly atypical spindle and giant cells with sarcomatous appearance. (C) 400x large tumor giant cells with sarcoma-like appearance. (D) 400x vimentin positivity in tumor cells.

Disclosures

Author contributions: All the authors contributed to evaluating and managing the case and to writing the manuscript. GR Gondu is the article guarantor.

Financial disclosure: No conflicts of interest or funding sources to disclose.

Informed consent was obtained for this case report.

Received: September 19, 2013; Accepted: December 17, 2013