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Received May 27, 2008;
 Accepted after revision January 26,
 2009.

Iran J Radiol 2009;6(2):93-96

Pure Yolk Sac (Endodermal Sinus) Tumor of the Liver: A Case Report

Hepatic tumors are rare in children. About 50%- 60% of these tumors are malignant. 65% of the liver malignancies are hepatoblastomas and most of the remainder are hepatocellular carcinomas. Yolk sac tumor (YST) is an extremely rare tumor of the liver.

We report on a hepatic yolk sac tumor in a 15-month-old girl who presented with acute abdomen. She was diagnosed initially as intussusception, while ultrasonography and CT-scan indicated a liver mass. Finally, yolk sac tumor was diagnosed surgically and histopathologically.

Keywords: Endodermal Sinus Tumor, Liver, Yolk Sac, Ultrasonography, Tomography, X-ray Computed

Introduction

Hepatic tumors are rare in children. These tumors are malignant in 50%-60% of the cases. Most of them are hepatoblastoma followed by hepatocellular carcinoma. Other rare malignancies are angiosarcomas, rhabdomyosarcomas, undifferentiated sarcomas and malignant germ cell tumors.¹ Yolk sac tumor (YST), rare but highly malignant, is the most common type of gonadal tumor in childhood.² It typically arises from the testes or ovaries. It is also known as endodermal sinus or Teilm tumor. There are two histological patterns. One is a pure YST; the other is associated with teratoma, dermoid cyst or choriocarcinoma.^{3,4}

YST is extragonadal in 10-15% of the patients.¹ This tumor is located mostly in the sacrococcygeal region.⁵ Other less common anatomic locations include the mediastinum and the central nervous system.^{6,7} YSTs have also been reported in the gastrointestinal tract, liver, nasal sinuses, uterus, cervix, thyroid, heart and diaphragm.⁸

YST of the liver is exceedingly rare and was first reported in 1975 in an 18-month-old boy who died 6.5 months after an extended hepatectomy.⁶ The second YST was reported in the falciform ligament.⁹ Although less than 20 mixed YSTs have been reported so far, few of them have been pure abdominal YSTs. According to a study published by Lacy et al in 2006, vaginal YST and pure liver YST have not been reported during the recent ten years.¹⁰

Case Presentation

A 15-month-old girl was admitted to Tabriz Pediatric Hospital with abdominal pain, vomiting and a low-grade fever. She had a one-month history of intermittent abdominal discomfort, loss of appetite and constipation. A palpable mass was revealed during physical examination. Vital signs and laboratory findings are shown in Tables 1 and 2.

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Table 1. Vital Signs

Blood Pressure	Pulse Rate	Respiratory Rate	Body Temperature
100/70	97/min	30/min	38.2 c

Table 2. Initial Laboratory Parameters

White blood cell	Hemoglobin	Hematocrit	Platelets	BUN,Cr,Na,FBS,U/A
14000/ μ l	10.3 gr/L	30.6%	338000	Normal Range

Table 3. Liver function Tests and Serologic Tests

Total Bilirubin	Direct Bilirubin	ALK (64-306)	LDH (185-640)	SGOT (5-40)	SGPT (5-40)	AFP (<10ng/ml)	β -hCG (<10IU/ml)
0.9 mg/dl	0.2 mg/dl	387	889 U/L	95 U/L	10 U/L	338 ng/ml	0.89 IU/ml

After undergoing ultrasonography (ULTRASONIX, OP, Canada) intussusception was ruled out. Hepatomegaly (the liver span was 95mm, while the normal range for children under 2 years is 70-90mm) and a well defined mixed echogenic mass (65×58×56mm) with central necrosis in the right liver lobe were reported (Figs. 1. A&B).

After encountering liver mass, liver function tests and serologic tests were carried out (Table 3).

CT-scan (BALANCE SIEMENS, Germany) demonstrated a heterogeneous mass (65×65mm) with well-defined margins and central necrosis in the antero-inferior segment of the right lobe of the liver (Fig. 1C). It was near to the anterior abdominal wall with

no mass effect on the intestinal loops.

During surgery, a mass in the inferio-anterior segment of the right lobe (6×6 cm), separable from the parenchyma was reported. It was fragile and probably related to Glisson's capsule and adhered to the anterior abdominal wall. It was resected completely.

The specimen was evaluated by two pathologists. It was a soft, fragile, yellow-gray tissue containing multiple cysts. Sections showed loose meshwork of spaces lined by primitive tumor cells with hyperchromatic, irregular, large nuclei containing prominent nucleoli and also glomerulus-like structures composed of a central blood vessel, enveloped by neoplastic cells within a space similarly lined by the same type of

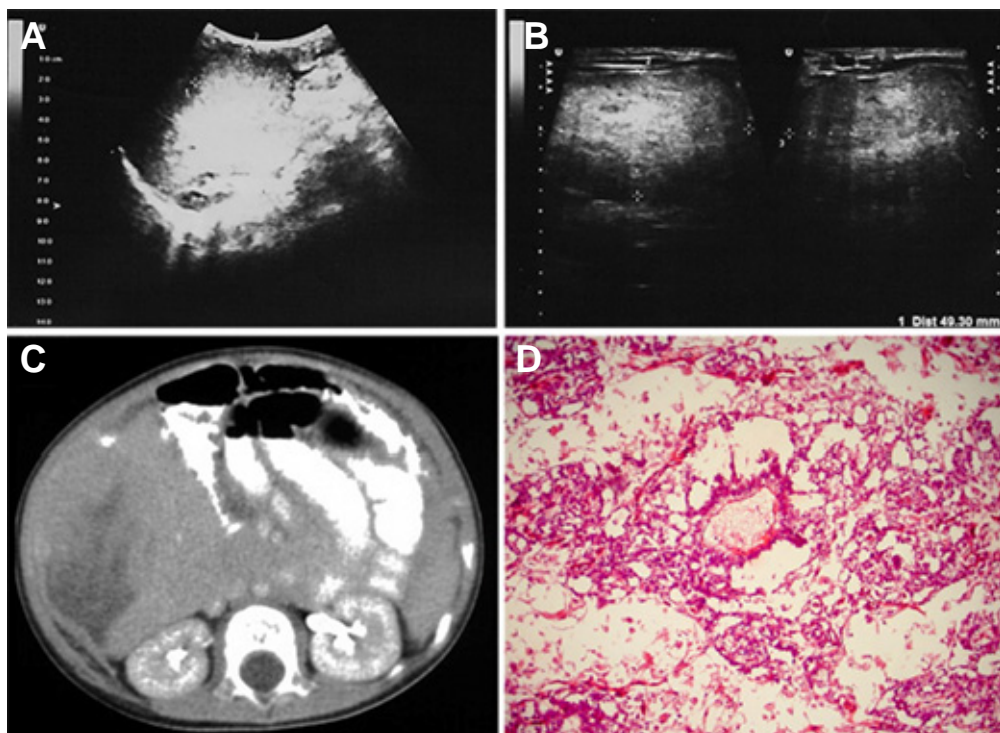


Fig. 1. A 15-month-old girl with pure yolk sac tumor of the liver. **A&B.** Longitudinal scan of ultrasonography demonstrated a well-defined mass in the antero-inferior segment of the right liver lobe. **C.** Axial CT scan enhanced with IV and oral contrast shows a well-defined solid mass with central necrosis localized in the antero-inferior segment of the right lobe of the liver with a faint heterogeneous enhancement. **D.** Histopathological section of the resected mass shows a characteristic large Schiller-Duval body structure that simulates a renal glomerulus structure.

primitive cells (Schiller-Duval bodies) (Fig. 1D).

The patient was discharged in good condition and received chemotherapy. Relapse or complications were not seen during 3, 6, 9 and 12-month follow ups.

Discussion

YST accounts for less than 1% of germ cell tumors.^{11,12} An extragonadal location is unusual and the tumor is generally located in the mediastinum, pineal gland, sacrococcygeal region, and vagina.^{3,4} In the ten recent years, few pure YSTs have been reported in the abdominal region in children.¹⁰ To our knowledge, only two pediatric cases of pure YSTs have been reported in the liver in English literature.^{6,9}

Differential diagnoses of hepatic malignancies are hepatoblastoma, hepatocellular carcinoma, angiosarcoma, rhabdomyosarcoma, undifferentiated sarcoma and malignant germ cell tumor.¹¹ Differentiation of each of the above diagnoses is impossible by imaging methods without excisional biopsy.¹³

The main clinical presentations in the reported cases were chronic upper abdominal pain and weight loss, abdominal distension without acute abdomen or intestinal obstruction.¹³ Abdominal discomfort was experienced in the present case as well, but the chief complaint was acute abdomen.

Alpha-fetoprotein (AFP) and α_1 -antitrypsin almost always rise in YSTs but β -HCG and carcinoembryonic antigen (CEA) remain normal.³ Mild elevation of the liver enzymes have been reported, however the diagnostic value of these tests are limited.¹ In this case, liver enzymes and β -HCG were normal, while the AFP level had 33 folds elevation.

The histopathological study of the present case demonstrates Schiller-Duval bodies, which are the hallmark of YST.¹⁴ The following wide range of histopathological patterns may be observed in YST: 1. Microcystic, 2. Endodermal sinus, 3. Solid, 4. Alveolar glandular, 5. Polyvesicular, 6. Myxomatous, 7. Papillary, 8. Macrocystic, 9. Hepatoid and 10. Intestinal.¹⁵ Frequently the radiological presentation is like a solid mass.

YST is a poor prognosis malignancy with and the treatment is lobectomy or liver transplantation and chemotherapy. Previous reported cases were treated

with resection and chemotherapy and only two unresectable cases (2-year-old boys) were treated successfully with liver transplantation.^{12,16} The tumor was successfully resected and followed up by effective chemotherapy with no recurrence in the present case.

We conclude that if a liver mass with a solid or complex appearance, is encountered in children, particularly in the presence of a normal level of β -HCG and a significant elevation of AFP, yolk sac tumor should be considered; despite its rare incidence.

Acknowledgements

We would like to acknowledge Professor Saeed Rad and Dr. Nick Alapour for their comments.

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