VIA MEDICA

CASE REPORT

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A case of a patient with embryonal sarcoma presenting with abdominal pain

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ABSTRACT

Undifferentiated embryonal sarcoma of the liver is a rare and aggressive pediatric neoplasm. Due to its features in the imaging studies, there is a high rate of misdiagnoses. We present a 16-year-old female referred to our hospital with abdominal pain. At the initial work-up, we suspected a hydatid cyst as one of differential diagnoses due to the cystic pattern of the mass on the computed tomography scan. The needle biopsy smear was sent for pathology analysis which was negative for scolex of *Echinococcus granulosus*. However, the pathology report indicated neoplastic features in the biopsy. She underwent surgery and total resection was performed. The mass was sent for further investigation which confirmed the diagnosis of embryonal sarcoma with osteosarcomatous components. Embryonal sarcoma should be suspected in large tumors at any age.

Keywords: abdominal pain; sarcoma; case report

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Introduction

After hepatoblastoma and hepatocellular carcinoma, undifferentiated embryonal sarcoma (UES) is the third most prevalent primary malignant liver tumor in pediatrics. The majority of patients are between the ages of 6 and 10, without ethnic or sex predominance [1]. Stocker and Ishak described UES of the liver for the first time in 1978 [2]. It is mesenchymal in origin and rare in adults. Although UES is the third most frequent primary malignant tumor of the liver in the pediatric population, few cases have been reported in the literature. The presentation may include fever, weight loss, and pain. Additional signs and symptoms may include anorexia, vomiting, diarrhea, lethargy, constipation, and respiratory distress [1].

Here we report a case of a 16-year-old female who was admitted to our hospital with acute abdominal pain. We suspected the presence of a hydatid cyst as one of her differential diagnoses due to the cystic pattern of the mass on the computed tomography scan.

Case presentation

A 16-year-old girl was admitted to our hospital with complaints of progressive abdominal pain located in the right upper quadrant. It was her first presentation. On further inquiry, it turned out that she had no weight loss, was not febrile, or icteric. On physical examination, a huge, non-tender, and immobile liver was palpable in the right upper side of the abdomen. No palpable lymphadenopathy was detected, and her vital signs were within normal ranges. Laboratory tests showed normal liver function tests. Further examination was done. Computed tomography (CT) showed a well-defined low-density large heterogenous cystic lesion (190 × 115 × 163 mm) with solid components and

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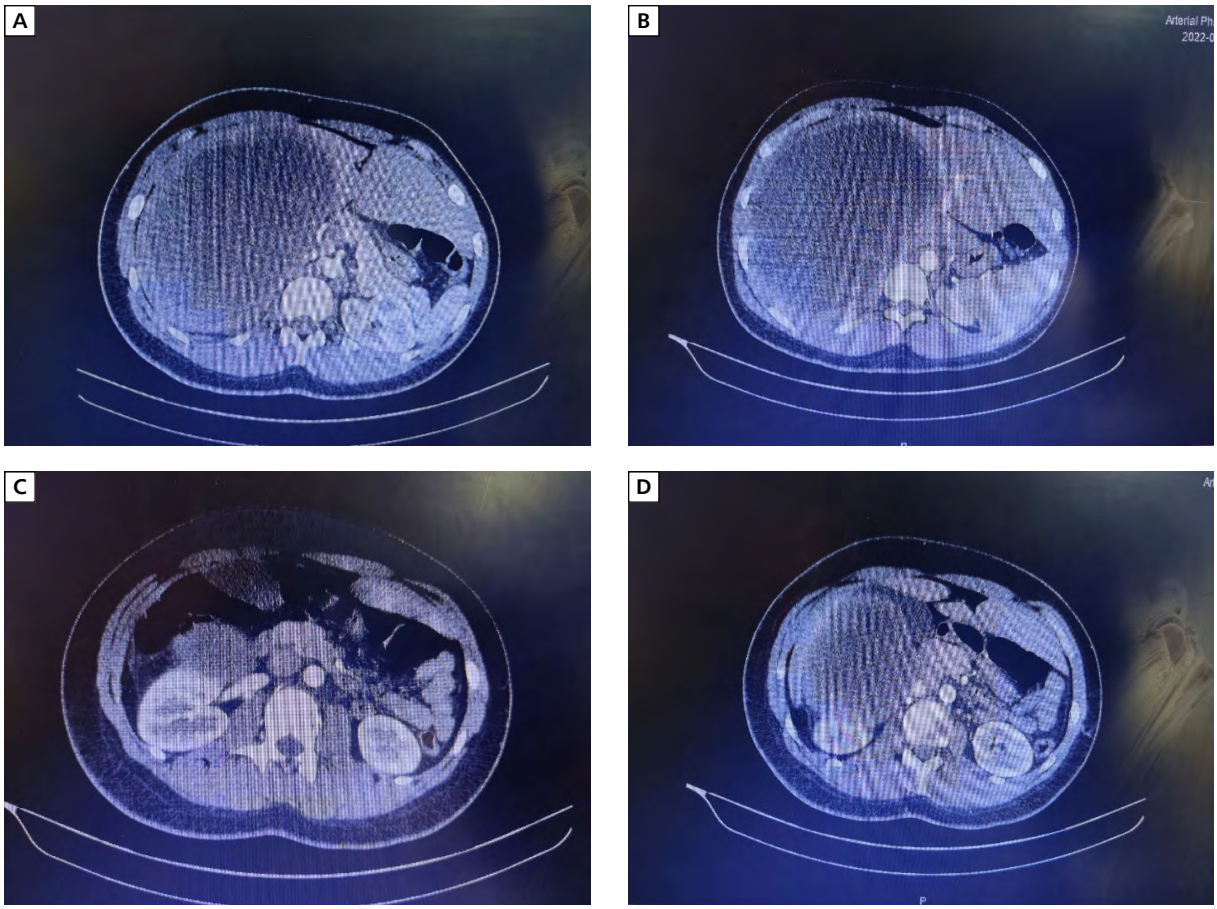

Figure 1A–D. Computed tomography scan showing a massive lesion in the liver: a well-defined low-density large heterogenous cystic lesion

enhanced septa in the right lobe of the liver (Fig. 1). On sonography, a huge solid cystic mass was seen in the right lobe of the liver. This massive mass had compressive effects on surrounding structures including the portal vein. Due to suspicion of hydatid cyst, the needle biopsy smear was sent for pathology examination which turned out to be negative for scolex of *Echinococcus granulosus.*

The biopsy pathology report indicated embryonal sarcoma with osteosarcomatous components. She underwent surgery with total resection of the tumor with nearly 2 cm tumor-free margins (Fig. 2). The right lobe of the liver, right biliary duct, and distal part of the duct were resected completely and sent for further histopathology investigation. It should be noted that due to the size of the mass and its highly compressive effect, the medical team decided that surgery was the best strategy because it seemed that chemotherapy would not be beneficial given the size of the mass.

Histopathology review documented malignant spindle cells and oval cells set in myoxid stroma. The cells had a high N/C ratio and hyperchromatic nuclei. Central necrosis and osteoid formation were also identified. On immunohistochemistry, CD10 was positive and SMA weakly scattered positive. After surgery, the patient was stable and was discharged with a follow-up treatment plan. She was followed up regularly, with no signs of recurrence on the last visit 10 months after surgery.

Discussion

Embryonal sarcoma of the liver is an aggressive mesenchymal tumor that occurs predominantly in pediatric patients. Despite years of research, the pathophysiology of this condition is still unknown. The prognosis for patients has been significantly improved by multimodal therapy, which includes surgery, chemotherapy, and radiation therapy. For better results, this successful management requires early diagnosis.

There are many possible diagnoses for undifferentiated liver embryonal sarcoma. Since each liver disease occurs in a certain age range, the patient's age

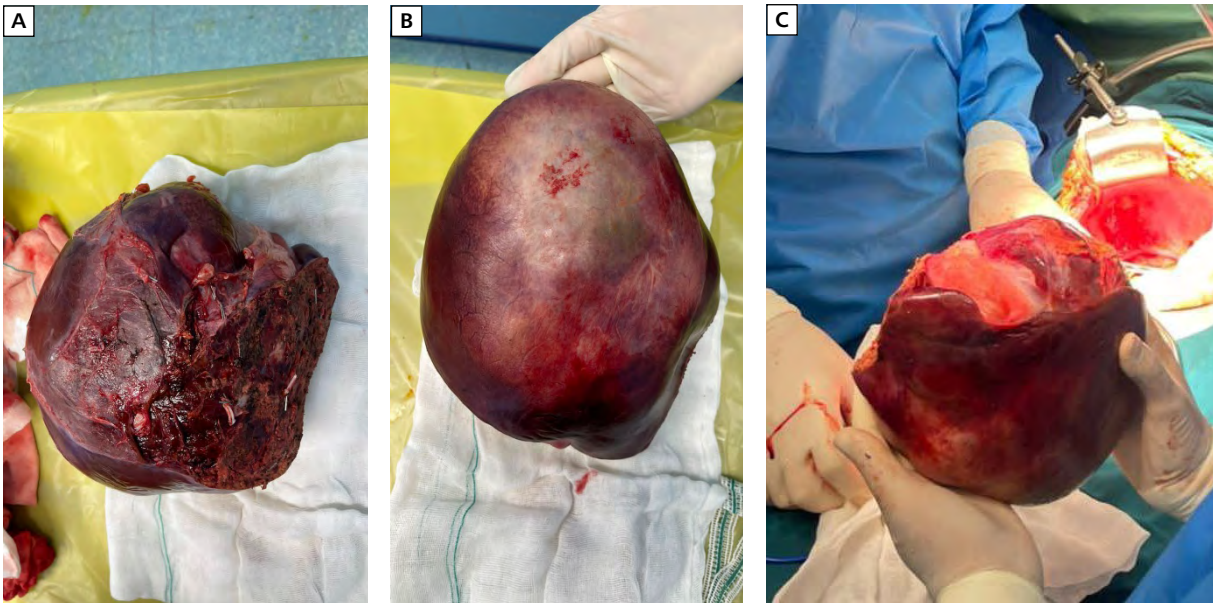

Figure 2A–C. The resected tumor

is frequently useful in reducing the number of possible differential diagnoses. However, our patient was older than the usual onset age. The UES of the liver is most often diagnosed in patients aged from 6 to 10 years.

In 2017, an 8-year-old male [3] had unsuccessful surgery for presumed hydatid disease which finally turned out to be UES of the liver located in the right lobe as in the case of our patient. Yoon et al. also reported a case of UES of the liver that was incidentally found in a 53-year-old female, which at first raised suspicion of a hydatid cyst. The follow-up CT scan suggested a neoplastic mass rather than a simple cyst [4].

Undifferentiated embryonal sarcoma of the liver is more frequent in the right lobe of the liver than in the left lobe [5]. After neuroblastoma and Wilms tumor, primary hepatic tumors are the third most common solid excrescences in pediatrics, accounting for approximately 2 percent of all pediatric cancers. Malignant mesenchymal hepatic tumors, hepatoblastoma and hepatocellular carcinoma, although rare, are very important in pediatrics [6].

The clinical features of UES are not specific. The signs and symptoms are usually related to the mass and its compressive effects on surrounding structures, as shown in this case. Palpable abdominal mass with or without upper abdominal pain may be found in some cases. Fever, which is found in the majority of tumors due to necrosis, hemorrhage, and cytokines effects, is not specific [7]. Undifferentiated embryonal sarcoma is not caused by cirrhosis or other chronic liver diseases; therefore, liver function tests and tumor markers including AFP, CEA, and CA19-9 are within normal limits in most cases.

Undifferentiated embryonal sarcoma is strongly positive for vimentin and 1-antitrypsin and focally positive for cytokeratin, desmin, α-SMA, muscle-specific actin, CD68, myoglobin, neuron-specific enolase, S100, and CD34, which suggests that an embryonic sarcoma is undifferentiated.

In 2020, Zhang et al. [8] reviewed retrospectively all patients referred to the Shenging Hospital from 2005 to 2017 and recruited 14 patients aged 2 to 60. They indicated that the preoperative imaging had a high misdiagnosis rate, and total resection was the first treatment choice, as in our patient.

According to Techavichit et al. [9], total resection of the tumor mass combined with neoadjuvant chemotherapy with ifosfamide and doxorubicin, cyclophosphamide plus doxorubicin plus vincristine, or ifosfamide plus etoposide showed better survival outcomes in the case of localized resectable cancers. Furthermore, May et al. [10] recommended adjuvant chemotherapy as an alternative for these cases, using vincristine, actinomycin D, and cyclophosphamide (VAC) regimens. For patients with unresectable or advanced tumors, Techavichit et al. [9] recommended liver transplantation.

Undifferentiated embryonal sarcoma prognosis varies highly with survival ranging from 20 to 100 percent [9]. Techavichit et al. [9] and Zhang et al. [11] demonstrated that complete tumor resection is the key factor

in increasing the survival rate of patients with resectable UES tumors.

Due to the low prevalence of UES, misdiagnoses, such as hepatic abscess, hemorrhage cystic tumor, and hydatid cyst, are common [12].

Conclusions

Undifferentiated embryonal sarcoma should be considered in differential diagnosis of large liver tumors regardless of patients' age. Our case shows that early surgery can have the same results as a combination of chemotherapy and surgery to secure a better survival rate.

Article Information and Declarations

Ethics statement

All the performed procedures were in accordance with the ethical guidelines of Iran University of Medical Sciences and the Declaration of Helsinki 1975 (year 2008).

Written informed consent was obtained from the studied patient.

Author contributions

All authors were responsible for designing the study, collecting data, and writing the article.

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None.

Conflict of interest

The authors declare that there is no conflict of interest.

Supplementary material

None.

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