



## Undifferentiated Embryonal Liver Sarcoma Diagnosed after Blunt Abdomen Trauma: A Case Report

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### Abstract

Undifferentiated Embryonal Liver Sarcoma (UELS) accounts about 13% of all malignant liver tumors in children. We present a case of 7-year-old boy with a large hepatic mass diagnosed incidentally after a blunt abdominal trauma. An extended right hepatectomy was performed. The postoperative pathology showed an UELS. The patient received postoperative chemotherapy. After more one year the computed tomography showed no evidence of recurrence. UELS is a potential treatable malignancy when treated with chemotherapy after complete surgical resection.

**Keywords:** Undifferentiated embryonal sarcoma; Trauma; Liver; Boy; Syria

### Introduction

Undifferentiated Embryonal Liver Sarcoma (UELS) is a highly malignant mesenchymal tumor. It was termed as UELS with a median survival of less than 1 year following diagnosis by Stocker, et al. [1]. It is the fourth childhood liver tumor (after hepatoblastoma, infantile hemangioendothelioma and liver cancer) [2]. UELS represents about 5-13% of all pediatric hepatic tumors [3]. This paper presents a case of a UELS mimicking a liver hematoma in its initial clinic-radiological presentation.

### Case Presentation

A 7-year-old boy was admitted to the Al Assad University Hospital in Damascus-Syria in June 2016. He presented with a three-week history of pain in the right hypochondriac region after a blunt trauma. He had neither history of hepatitis viral infections nor prenatal exposure to Phenytoin. A physical examination revealed a local tenderness in addition to gross hepatomegaly. Laboratory investigations demonstrated mild anemia (Hb 8g/dl, normal range: 12-14 g/dl). Liver test results and neoplastic markers were normal. Ultrasonography showed a large mass with mixed solid and cystic components. A subsequent contrast-enhanced abdominal computed tomography (CT) scan demonstrated an 8 cm × 13 cm multiloculated, complex cystic mass in the right liver lobe (Figure 1).

The initial impression was hematoma after liver rupture (caused by the trauma), however the conflicting between the findings of US (mass more than cyste) and the Ct (cyste more than mass) put the diagnosis of an undifferentiated embryonal liver sarcoma (UELS) at first. Subsequently, the patient underwent an extended right hepatectomy (Figure 2) on June 9, 2016. Intraoperatively, a huge mass was revealed. Grossly, partial hepatectomy was performed; the specimen measured 16 cm × 14 cm × 7 cm, and weighed 540 gram (Figure 3). The tumor measured 9 cm × 8 cm × 6 cm. It was fleshy soft, and gray in color with areas of necrosis and hemorrhage. The obtained sections showed proliferation of highly atypical pleomorphic cells with frequent mitoses. Diagnosis of high grade malignancy; was applied. Microscopic examination revealed cellular proliferation within myxoid stroma, composed of pleomorphic cells (epithelioid and spindle cells, and numerous multinucleated cells) having large pleomorphic atypical nuclei (Figure 4), and showing prominent mitotic activity with atypical mitotic figures (Figure 5). Foci of necrosis and hemorrhage were observed (Figure 6). The tumor cells included eosinophilic globules that showed positive staining on PAS, and diastase-PAS. Immunohistochemistry negative result was on AE1/AE3, HepPar1, AFP, Myogenin, anti-CD34, anti-CD56, and antivimentin. MIB1 showed high index. Postoperatively, the patient developed a bile leak, which was treated well by Endoscopic Retrograde Cholangiography (ERC) and stenting. The boy was discharged 13 days after the operation. A six months course of chemotherapy is including vincristine (V), doxorubicin (D), cyclophosphamid (C), ifosfamide (I) and etoposide (E) in VDC/IE protocol was initiated, starting at 4 weeks after the operation, and was well tolerated by the boy. Recently, he is clinically disease-free, 15 months after resection. A follow-up CT scan performed 15 months after the operation did not reveal residual or metastatic disease.

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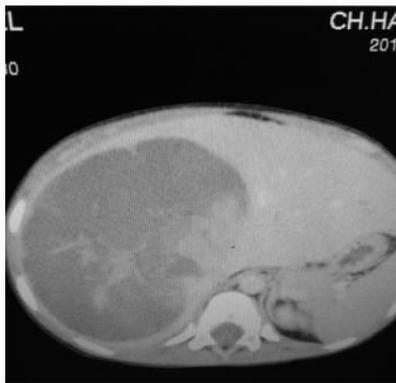
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**Figure 1:** CT scan of the abdomen shows a huge mixed cyst/mass in the right hepatic lobe.



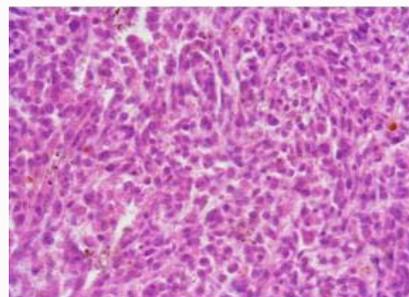
**Figure 2:** The resected surface following extended right hemihepatectomy.



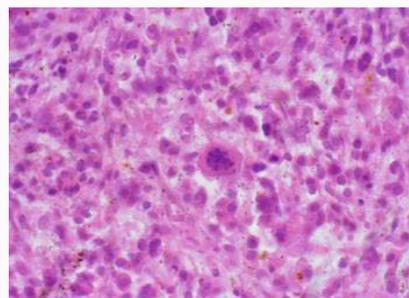
**Figure 3:** The resected tumor (Liver segments IV, V, VI, VII & VIII).

## Discussion

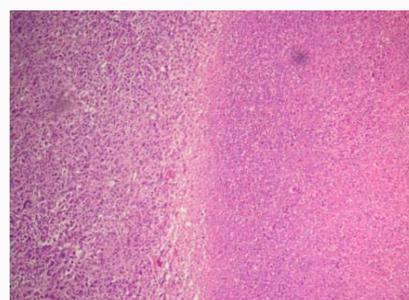
UESL is a rare hepatic mesenchymal tumor that was first reported and classified by Stocker, et al. [1]. UESL occurs in children as well as in adult, with a peak incidence between 6 and 10 years of age, without gender predilection [4-6]. Our patient presented with a painful hepatomegaly after a blunt trauma. The literature showed, that patients with this entity present with a variety of nonspecific symptoms and signs such as nausea, vomiting, upper abdominal pain, weight loss, fatigue, jaundice and palpable abdominal mass [7-9]. While our case was diagnosed after a blunt trauma, there are some colleagues reported some cases of intraperitoneal hemorrhage due to spontaneous rupture of the tumor [10-13]. Many studies, inclusive our case, reported that laboratory findings are non-specific and the tumor markers are not increased [14]. Some cases demonstrated an elevation in serum Alkaline Phosphatase (ALP) and lactate dehydrogenase



**Figure 4:** Dense cellular area, of large pleomorphic atypical cells.



**Figure 5:** Atypical mitotic figures.



**Figure 6:** The tumor necrosis.

(LDH) [8]. Evaluation of some tumor markers including Alpha Feto Protein (AFP), and Carcinoembryonic Antigen (CEA) is noted in other cases [15,6]. While many UESL are mistakenly diagnosed as a hematoma, hydatid cysts or pyogenic abscesses [16-19], the conflict between the ultrasound (irregular hypo- or hyperechoic tissue) and CT findings (cystic changes) remains the key of the diagnosis. For unclear reasons, the tumor is mainly localized in the hepatic right lobe. UESL typically has a huge size [20]. According to many studies, the combination of complete resection and chemotherapy with or without radiation remains the cornerstone for long survival rates [19-23]. Additionally, other authors demonstrated that UESL patients who underwent Transarterial Chemoembolization (TACE) followed by complete resection exhibited good results [20,24]. Kelly and Plant demonstrated that hepatic transplantation is an effective therapeutic method for UESL patients under certain circumstances [25,26]. More and more literatures reported the tumor is potentially treatable [24,27,28]. After fifteen months, our patient is in a very well physical status and there is no evidence of local or metastasis.

## Conclusion

Several points are worth to be recommended. Firstly, every cystic liver mass showed by CT scan, which looks like as tissue mass by

US is highly suspected to be malignant hepatic tumor, especially in childhood. At second, that suspected mass needs no biopsy as well as it is resectable. Thirdly, the combination between the chemotherapy and complete resection provides a good chance for long-term survival. A last, a multidisciplinary approach is always necessary.

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