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CASE REPORT

# Hanging Undifferentiated Embryonal Sarcoma of the Liver in Adult: an Unusual Presentation of an Aggressive Tumor

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## Introduction

Undifferentiated embryonal sarcoma of the liver (UESL) is a rare, highly aggressive hepatic neoplasm of mesenchymal origin seen almost exclusively in pediatric population [1]. It was termed UESL for the first time by Stocker et al. in 1978 [2]. This disease is very rare in adults with less than 70 cases reported in English literature [3–5]. Though the tumor usually presents as a large liver tumor in an adult, a giant (> 10 cm) and hanging tumor arising from the Reidel's lobe of the liver over a narrow stalk is very unusual and has not been reported in literature to date. We describe a case of a 29-year-old young female with a short clinical course and resectable hanging UESL, but unfortunately developed early recurrence despite R0 resection, signifying a poor prognosis disease.

## Case Report

A 29-year-old lady with no comorbid illness presented with 15 days history of pain and fullness in the right upper abdomen. She denied any history of jaundice, fever, or altered bowel habit. Her general physical examinations were normal. An abdominal examination revealed a large (15 × 12 cm), well-defined, nontender, firm lump in the right hypochondrium reaching up to the right iliac fossa. The lump was moving with respiration with a side to side mobility. Her blood investigations including hemoglobin and liver and renal function tests were within reference range. Contrast-enhanced computed tomography (CECT) of the abdomen showed a well-defined, (18 × 12 cm) homogenously enhancing mass lesion arising from the Reidel's lobe of the liver in a pedunculated fashion over a narrow stalk (Fig. 1). The tumor markers: serum alpha fetoprotein and carcinoembryonic antigen were normal. The preoperative fine-needle aspiration cytology showed spindle-shaped cells consistent with liver sarcoma. The diagnosis of resectable malignant liver tumor was made at the tumor board and planned for upfront surgery followed by adjuvant therapy. Intraoperatively, a large, well-marginated, purplish-blue, soft tumor was hanging from the Reidel's lobe, which was resected with a wide margin. Cut section of the resected specimen showed soft, spongy, solid-cystic lesion with hemorrhagic areas (Fig. 2). Postoperative period was uneventful and was discharged on the seventh postoperative day in a satisfactory condition.

Histopathology revealed proliferation of oval to spindle bizarre atypical cells arranged in sheets and scattered singly in a loose stroma. Tumor cells were pleomorphic with hyperchromatic nucleus and moderate to abundant cytoplasm. There were numerous bizarre-looking cells and tumor giant cells. Immunohistochemistry showed positivity for desmin (Fig. 3) and vimentin but was negative for cytokeratin, S-100, and HMB45. The microscopic resection margins were free of tumor. Based on the above characteristic description, diagnosis of undifferentiated embryonal sarcoma of the liver was made. She was advised for adjuvant combination chemotherapy at the earliest, but the patient and the supporting family member denied for it (because of financial constraints, cost, toxicity, and chances of recurrence in spite of chemotherapy),

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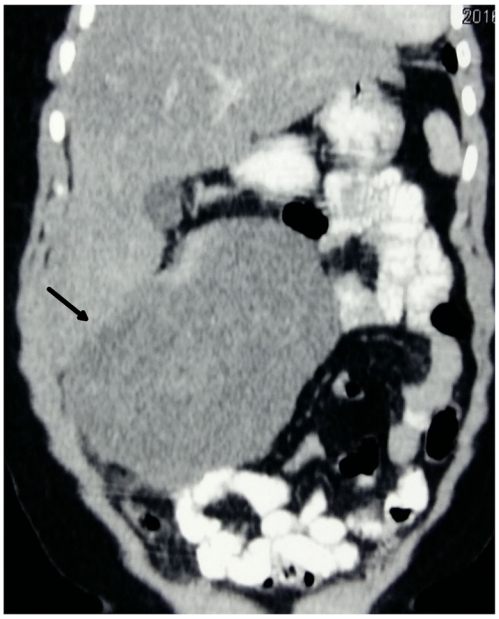

**Fig. 1** Contrast CT showing well-defined, pedunculated, homogenously enhancing mass lesion arising from the Reidel's lobe (arrow).

in spite of our preoperative counseling regarding its necessity. At 4 months of follow-up, she unfortunately developed diffuse peritoneal and liver recurrence and succumbed at 8 months of surgery.

## Discussion

Undifferentiated embryonal sarcoma of the liver is the third most common malignant hepatic neoplasm of pediatric population. Most often, it occurs in late childhood (age group of 6 to 10 years) and is infrequently seen in adults [1, 4]. Even in adults, most of the reported cases have occurred within 30 years with female preponderance and comprise less than 1% of sarcomas [6]. The tumor is usually asymptomatic to begin with until it acquires a large size. The symptoms initially are usually nonspecific. They usually present with large palpable mass with or without abdominal pain, weight loss, nausea or vomiting, and rarely with spontaneous tumor rupture and hemorrhage [7–9]. The duration of illness is usually short and acquires a large size because of its rapid tumor growth as seen in our case.

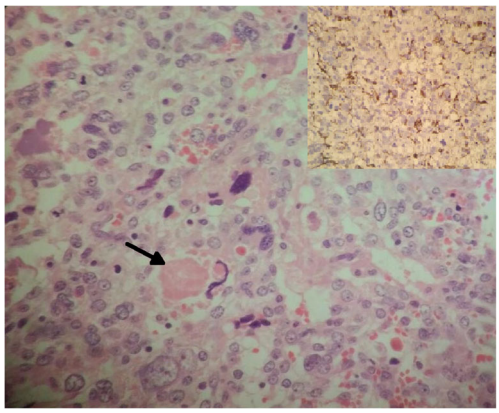

**Fig. 3** High-power microscopy (H&E, × 100) showing round to spindle bizarre pleomorphic tumor cells along with tumor giant cells (arrow)

Histopathologically, tumor cells appear as pleomorphic spindle or stellate shaped, within myxoid matrix with presence of intracytoplasmic eosinophilic granules and bizarre giant cells. Immunohistochemical studies show expression to vimentin, without staining for epithelial markers [1, 10].

Diagnosing an UESL in an adult is a difficult task because of its non-specific presentation, its short duration history, and

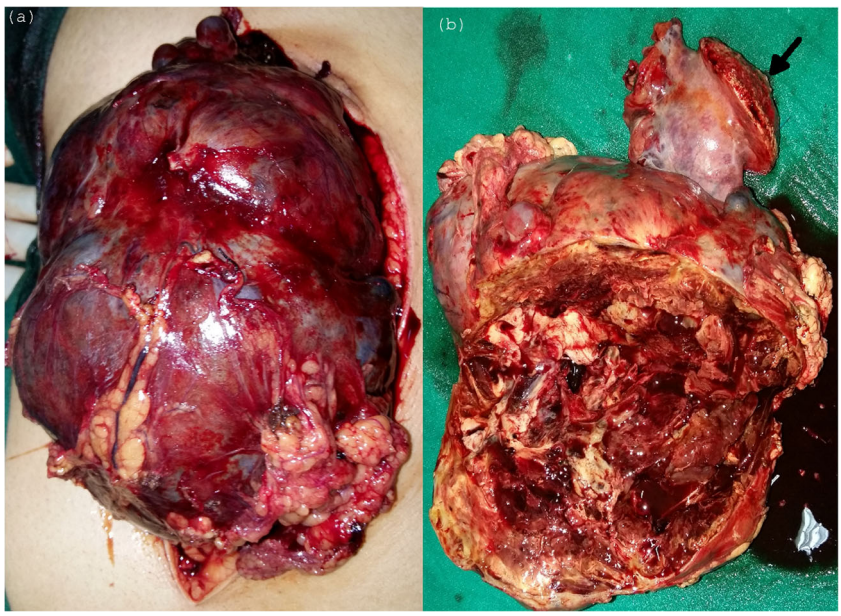


**Fig. 2** Intraoperative (**a**) and cut section (**b**) of resected tumor showing soft, spongy solid-cystic areas with a variegated appearance over a narrow pedicle (arrow)

its rare entity. However, the diagnosis of primary liver sarcoma should be suspected whenever any young patient presents with non-specific symptoms and a large liver mass on CT abdomen [11]. The liver mass appears solid on sonography and cystic appearance on CT, making the findings with discrepancy, which should raise the suspicion for this tumor. Moreover, the tumor is well-defined, heterogenous with solid-cystic appearance with normal tumor marker level [12]. Sometimes, the disease may mimic a benign cystic tumor (hydatid liver disease) making a diagnosis delayed [13, 14]. Unlike hanging hepatocellular carcinoma (HCC), UESL can also present as hanging tumor of the liver and should be kept as one of the differentials.

Although there is no consensus on the optimal management of this disease due to its rarity, it appears that the best treatment strategy is complete surgical resection of the tumor with R0 resection margin, which often requires major hepatectomy [15, 16]. But even after complete resection with negative microscopic margin, majority of patients die of tumor recurrence or metastases early after surgery, signifying aggressive tumor biology (Table 1). Therefore, multimodality treatment of neoadjuvant chemotherapy (any combination of cisplatin, cyclophosphamide, doxorubicin, vincristine, or ifosfamide) and adjuvant chemotherapy should be added to surgical resection to address micrometastases and improve survival based on the little published experiences [15, 17, 20].

The prognosis of UESL in adults was poor until recently when improvement in the survival was achieved by surgical resection and combination chemotherapy in neoadjuvant and adjuvant setting [9, 18]. Significantly better survival has been observed for patients receiving adjuvant chemotherapy as opposed to surgery alone [19] (Table 1). In one study, 42% of patients undergoing complete resection without adjuvant therapy recurred at 8 months compared to 23% at 28 months. In a study by Cao et al. [6], nine cases of UESL were studied and the follow-up data showed four of the nine cases had recurrence, and two patients died. Time to recurrence in those cases was 19, 4, 29, and 14 months. The mean OS was 58.25 ± 9.1 months after receiving adjuvant chemotherapy. They also saw the early recurrence after R0 resection at 4 months as was seen in our case. Similarly, in a review of 67 adult patients with UESL by Lenze F et al. [4], the median survival was 29 months and survival was longest in patients who underwent complete tumor resection and adjuvant chemotherapy. The actual 1- and 2-year survival rate for all patients was 61 and 55%, respectively.

In our case, although the tumor was hanging from the Reidel's lobe over a narrow pedicle and was completely excised with negative margin, she unfortunately developed early recurrence at 4 months pointing towards the aggressive tumor biology.

**Table 1** Summary of reported outcomes of 10 patients with UESL in adults

| Author | Year | Age/sex | Symptoms | Maximum size of tumor (cm) | Treatment | Chemotherapy | Recurrence | Follow-up (months) |
|---|---|---|---|---|---|---|---|---|
| Khan ZH [15] | 2017 | 21/M | Abdominal pain | 13 | OLT | Yes (neoadjuvant) | No | 18 (alive) |
| Mori A [17] | 2017 | 65/F | Backache | 16 | Right trisegmentectomy | No | 18 months | 26 (dead) |
| Treitl D [16] | 2016 | 30/M | Abdominal pain, early satiety | 45 | Bland embolization, left hespatectomy | Yes (adjuvant) | No | 3 (alive) |
| Zanwar S [9] | 2016 | 20/M | Abdominal pain | NA (ruptured liver tumor) | Right hepatectomy | Yes (neoadjuvant + adjuvant) | No | 24 (alive) |
| Giakoustidis DE [18] | 2016 | 30/M | Abdominal pain, fever | NA (voluminous tumor) | Right portal vein embolization, right trisegmentectomy | Yes (adjuvant) | 12 months | 28 (dead) |
| Hong WJ [11] | 2014 | 67/M | Abdominal pain | 13 | Segmentectomy (5, 6) | Yes (palliative) | 1 month | 8 (dead) |
| Chen JH [10] | 2013 | 63/M | Abdominal pain | 27 | Left lateral segmentectomy | No | 9 months | 12 (dead) |
| Jia C [19] | 2013 | 46/F | Abdominal pain | 6 | Segmentectomy (5, 4a) | No | 6 months | 12 (dead) |
| Lightfoot N [7] | 2012 | 78/F | Abdominal mass | 16 | Right hepatectomy | No | No | 6 (alive) |
| Kim HH [14] | 2011 | 47/F | Abdominal mass | 12 | Left lateral segmentectomy | Yes (adjuvant) | Yes (no time duration) | 48 (dead) |

*OLT;* orthotopic liver transplantation, *NA;* not available

## Conclusion

UESL is a rare tumor in adults with poor prognosis. Rarely, it can arise as a hanging tumor from the Reidel's lobe of the liver. Increased survival and prevention of recurrences can probably be achieved by surgical resection of the tumor combined with neoadjuvant and adjuvant chemotherapy.

### Compliance with Ethical Standards

**Conflict of Interest** The authors declare that they have no conflict of interests.

**Consent** Informed consent was obtained.

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