Julia Costas Eimil*, Paula Sánchez-Sobrino, Olaia Díaz Trastoy

*Servicio de Endocrinología y Nutrición, Complejo Hospitalario Universitario de Pontevedra, Pontevedra, Spain*

*Corresponding author.
*E-mail address:* julia.costas.eimil@sergas.es (J. Costas Eimil).

2444-3824/ 

# Hepatic undifferentiated embryonal sarcoma in a young adult

## Sarcoma embrionario indiferenciado hepático en adulto joven

Undifferentiated embryonal sarcoma of the liver (UESL) is an aggressive cancer with a very low incidence (0.1%–2% of liver tumours).[1] Although it also occurs in adults, it is more common in childhood.[2] Accurate preoperative diagnosis is a challenge. Survival has improved with the current treatment of complete surgical resection and chemotherapy.[3] We present a new case of UESL and discuss the central aspects of this type of cancer.

This was a 20-year-old woman with no relevant previous medical history who presented with vomiting and a palpable mass in the right hypochondrium. Abdominal ultrasound (Fig. 1A) and magnetic resonance imaging (Fig. 1B) showed a 12.5 × 12 × 10 cm multilocular cystic lesion in the right lobe of her liver. There were no abnormalities in her complete blood count, liver function tests or tumour markers. The preoperative radiological diagnosis was biliary cystic neoplasm. Bisegmentectomy V–VI was performed (Fig. 1C). She was discharged 24 h later with no postoperative complications. The pathology study revealed a high-grade undifferentiated embryonal sarcoma with free surgical margins. Microscopically, large spindle-shaped sarcomatous epithelial cysts were identified with high mitotic activity and hyaline globules. Immunohistochemically we found expression for desmin with activity for MYOD1 and negativity for beta-catenin, P53, P16 and myogenin. The patient was started on chemotherapy with vincristine, actinomycin-D and cyclophosphamide. Over the follow-up carried out to date (six months) there have been no signs of recurrence.

UESL was first described by Stocker and Ishak in 1978.[2] It is the third most common primary hepatic malignancy in childhood after hepatoblastoma and hepatocellular carcinoma. In adults, it is usually seen in women in their thirties or forties.[1] No obvious aetiological factors have yet been found, suggesting that it derives from the malignant transformation of a mesenchymal hamartoma.[1] The most common location is the right lobe of the liver; as in our case, it is usually more than 10 cm in size.[4]

Clinical presentation is variable, ranging from asymptomatic to non-specific symptoms (fever, weight loss and gastrointestinal symptoms) to severe symptoms caused by tumour rupture or liver failure.[1,2] An abdominal mass may be palpable on physical examination, with or without associated abdominal pain. There are no typical laboratory data of UESL,[3] with liver function tests and tumour markers usually normal, but elevation of transaminases, sedimentation rate and leucocytosis may be found.[3]

The radiological findings of UESL are not specific. Ultrasound usually shows a large, benign-appearing cystic-solid mass.[3] Computed tomography shows a large multi-septated hypodense mass with central necrosis. Magnetic resonance imaging is particularly useful for detecting small multifocal nodules and assessing vascular-biliary structures and hilar lymphadenopathy. The presence of a hypointense border caused by the peri-tumour fibrous pseudocapsule, intratumour bleeding, and heterogeneous enhancement both in the periphery and in the solid tumour components on late T1-weighted images are suggestive of UESL.[2,5]

The differential diagnosis of UESL is complex. The age of the patient can be indicative. The main differential diagnoses in the paediatric population are mesenchymal hamartoma, hepatoblastoma and biliary embryonal rhabdomyosarcoma.[3] In adults, it includes hepatocellular carcinoma, biliary cystic neoplasms, gastrointestinal stromal tumour, angiomyolipoma, epithelioid haemangioendothelioma and malignant melanoma.[3] An accurate, early diagnosis is key to increasing the chances of long-term survival.

The definitive diagnosis of UESL is based on histopathological examination and immunohistochemical evaluation. Macroscopically, it usually involves a single, heterogeneous lesion with a cystic-solid appearance, greyish in colour, with areas of necrosis delimited by a fibrous pseudocapsule.[3] Microscopically, the pseudocapsule separates the lesion from the surrounding liver parenchyma.

The variable expression of histiocytic, muscular and epithelial markers suggests that it originates from primitive stem cells. Most are positive for vimentin, desmin, CD68, B2-cell lymphoma, α1-antitrypsin and CD10. The lack of expression for the hepatocyte paraffin-1 antibody differentiates it from hepatoblastoma and hepatocellular carcinoma.[1]

Complete surgical resection and chemotherapy are the mainstays of UESL treatment. The combination of surgery and chemotherapy has improved outcomes, increasing five-year survival from 40% in the 1980s to the current 70%.[1–5] If R0 resection does not seem possible at diagnosis, neoadjuvant chemotherapy can be started.[5] Negative resection margin, tumour size less than 15 cm, and combination therapy (surgery and chemotherapy) are independent prognostic factors.[3–5] The presence of extrahepatic disease does not seem to significantly impact the prognosis and should not be considered a contraindication for surgery.[4] Liver transplantation may be an alternative for the technically unresectable disease after neoadjuvant chemotherapy or recurrence.[3,5] Recurrences usually occur within two years after surgery, with a higher rate if the resection margins are positive.[3]

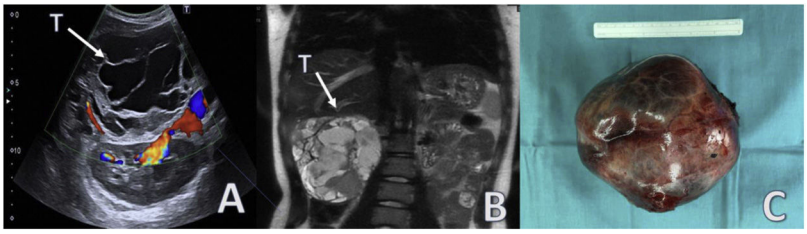


**Figure 1** A. Doppler ultrasound of abdomen/pelvis: multilocular cystic lesion in the liver with peripheral vascularity. T: tumour. 1B Magnetic resonance imaging. Cystic-solid multilocular lesion with hypointense border and heterogeneous enhancement in the periphery and within the solid components of the mass. T: tumour. 1C. Surgical specimen.

In conclusion, UESL is a very rare tumour, which is difficult to correctly diagnose preoperatively. Treatment is complete tumour excision and adjuvant chemotherapy.

## Funding

No funding was received for this study.

## Conflicts of interest

The authors declare that they have no conflicts of interest.

## References

1. Martins ACA, Costa Neto DCD, Silva JDDE, Moraes YM, LeÃo CS, Martins C. Adult primary liver sarcoma: systematic review. Rev Col Bras Cir. 2020;47:e20202647.
2. Meyers R, Hiyama E, Czauderna P, Tiao GM. Liver tumors in pediatric patients. Surg Oncol Clin N Am. 2021;30:253–74.
3. Shu B, Gong L, Huang X, Cao L, Yan Z, Yang S. Undifferentiated embryonal sarcoma of the liver in adults: retrospective analysis of a case series and systematic review. Oncol Lett. 2020;20:102.
4. Wu Z, Wei Y, Cai Z, Zhou Y. Long-term survival outcomes of undifferentiated embryonal sarcoma of the liver: a pooled analysis of 308 patients. ANZ J Surg. 2020;90:1615–20.
5. Techavichit P, Masand PM, Himes RW, Abbas R, Goss JA, Vasudevan SA, et al. Undifferentiated Embryonal Sarcoma of the Liver (UESL): a single-center experience and review of the literature. J Pediatr Hematol Oncol. 2016;38:261–8.

Lidia Betoret-Benavente[a,*], Gonzalo P. Rodríguez-Laiz[b,c], Paola Melgar-Requena[b,c], Joaquín Ruiz-López[b], Cándido F. Alcázar-López[b,c], Celia Villodre-Tudela[b,c], José Manuel Ramia-Ángel[b,c,d]

[a] *Servicio de Cirugía General, Hospital José María Morales Meseguer, Murcia, Spain*
[b] *Unidad de Cirugía Hepatobiliopancreática y Trasplante Hepático, Servicio de Cirugía General, Hospital General Universitario Dr. Balmis, Alicante, Spain*
[c] *Instituto de Investigación Sanitaria y Biomédica de Alicante (ISABIAL), Alicante, Spain*
[d] *Universidad Miguel Hernández, Alicante, Spain*

* Corresponding author.
*E-mail address:* lidiabetoret@gmail.com
(L. Betoret-Benavente).