Undifferentiated embryonal sarcoma of the liver - case report
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Section: Abdominal imaging
Area of Interest: Oncology Abdomen
Procedure: Diagnostic procedure
Imaging Technique: CT
Imaging Technique: Catheter arteriography
Imaging Technique: Ultrasound
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 17 years, female

Clinical History:
A 17-year old female presented with diffuse abdominal pain for the past few weeks. She had no relevant past medical history. There were no relevant physical findings and the laboratory assays were normal.

Imaging Findings:
Abdominal Computed Tomography (CT) revealed a well-circumscribed subcapsular lesion on the right lobe of the liver with solid and cystic components. The solid portion of the mass had lobulated morphology and soft tissue attenuation with rich vascularization, enhancing particularly in the portal phase. In the cystic component of the lesion, spontaneously dense areas that did not show enhancement could also be identified, corresponding to recent bleeding.

Abdominal ultrasound (US) showed the mass to be within the right lobe of the liver, large, heterogeneous and with a large cystic component.

Angiography revealed a predominantly avascular mass with smaller highly vascularized peripheral areas. These areas were vascularized predominantly by the right hepatic artery which was embolized with microspheres (size 900-1200?m) to reduce the risk of haemorrhage during surgery. Then right hepatectomy was performed, with resection of a well demarcated unruptured yellowish, roughly spherical, spongiform mass. Afterwards adjuvant chemotherapy and radiotherapy was administered.

Discussion:
Undifferentiated embryonal sarcoma (UES) is a rare type of malignant tumour of the liver, typically found in children and young adults. Nevertheless corresponds to the third most common liver tumour among the paediatric population [1].

UES is associated with poor prognosis but there are reports of long term survival after surgical resection and chemotherapy treatment [2]. Usually patients present with abdominal pain, palpable right upper quadrant mass, weight loss, jaundice or other gastrointestinal complaints. Serum ?-fetoprotein levels are not elevated [3].

UES typically presents as a large encapsulated solitary mass, often greater than 10 cm and more frequently in the
hepatic right lobe [1].

CT demonstrates a multicystic mass with septations and solid components [1, 4, 5]. Magnetic Resonance imaging (MRI) typically reveals an encapsulated hyperintense mass with hypointense septations on T2W images due to the presence of necrosis and mixoid material [5]. It can also demonstrate, on T1W imaging, haemorrhagic hypointense areas and a hypointense fibrous capsule [4, 5]. In general, most tumours do not exhibit enhancement, but the rim and internal septa have high attenuation. Ros et al. has reported that on angiography, half of the tumours were hypovascular, 17% were hypervascular, and 33% were avascular [6].

On CT and MRI this lesion has a cystic appearance because of the high water content of the abundant myxoid stroma. However on US this lesion is typically solid. This discrepancy between a cystic appearance on MRI and CT and a solid nature on US strongly suggests a diagnosis of UES [7].

The differential diagnosis depends on the features of the lesion. If a cystic or multicystic hepatic mass is found in a child, mesenchymal hamartoma, cystic hepatoblastoma or necrotic metastasis should be considered. If the mass is solid, hepatoblastoma, infantile hemangioendothelioma, hepatocellular carcinoma, and metastasis should be taken into account. In our case the tumour was subcapsular so other subcapsular lesions should be considered, including seeded metastasis (e.g. from an ovarian neoplasm).

UES are aggressive neoplasms with very poor prognosis. These tumours are typically large and difficult to resect, but distant metastasis is rare [1]. Imaging approach includes initial ultrasound with Doppler, followed by CT or MRI, to characterize the tumour and plan surgical resection. Surgical resection with neoadjuvant chemotherapy similar to that used for rhabdomyosarcoma offers the best long-term results and possibility of cure [2, 8]. In our patient, follow up imaging revealed residual tumour.

**Differential Diagnosis List:** Undifferentiated embryonal sarcoma of the liver, Mesenchymal hamartoma, Cystic hepatoblastoma, Necrotic metastase, Malignant fibrous histiocytoma, Rhabdomyosarcoma

**Final Diagnosis:** Undifferentiated embryonal sarcoma of the liver

**References:**


Description: Selective arteriogram of the common hepatic artery illustrating the hepatic parenchyma compressed by the lesion that has some vascularized solid components. Origin: Hospital de Santa Maria.
Description: Selective arteriogram of the right hepatic accessory artery emerging from the superior mesenteric artery. Origin: Hospital de Santa Maria
**Figure 3**

Description: Arterial phase CT illustrating the highly vascular solid components of the lesion. 

Origin: Hospital de Santa Maria
**Description:** Portal phase CT revealing a well-circumscribed hypoattenuating lesion with enhancing solid components. **Origin:** Hospital de Santa Maria
Figure 5

**Description:** Coronal reconstruction depicting the predominantly cystic right liver lesion

**Origin:**
Hospital de Santa Maria
Description: Axial plane of the hepatic right lobe illustrating that some areas of the cystic component present low level echoes, later corresponding to hemorrhagic areas on CT. Origin: Hospital de Santa Maria.
Figure 7

**Description:** Axial plane of the hepatic right lobe depicting a predominantly cystic lesion with a smaller solid component **Origin:** Hospital de Santa Maria
Figure 8

Description: Axial plane of the hepatic right lobe with color Doppler evaluation depicting the vascularization of the solid component of the lesion. Origin: Hospital de Santa Maria
Description: Unenhanced CT depicting a well circumscribed mass with cystic and solid components in the right lobe of the liver. Please note the spontaneously dense areas within the lesion corresponding to haemorrhagic component. Origin: Hospital de Santa María