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Case Report

Hepatic undifferentiated embryonal sarcoma as a cause of acute abdomen

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ABSTRACT

Primary liver sarcoma represents less than 1% of all primary liver tumors. Hepatic undifferentiated embryonal sarcoma (HUES) is included in the histologic group of primary liver sarcomas. HUES is a rare malignant liver tumor with a low incidence in the adult population. At moment there is no tumor marker that increases suspicion or diagnosis Early diagnosis is key to rise the possibilities of long-term survival. It has currently improved its prognosis through multidisciplinary management with surgical resection and chemotherapy. This case report presents a 20-year-old female patient with acute abdomen and anemic syndrome secondary to spontaneous rupture of a HUES requiring medical management initially based on resuscitation and arterial embolization with subsequent surgical resection.

Keywords: HUES, Hepatic bisegmentectomy, Hepato-splenicscintigraphy

INTRODUCTION

Primary liver sarcomas are rare and aggressive tumors with a poor outcome, representing less than 1% of primary liver tumors. Undifferentiated embryonal sarcoma represents 7% of histological lines of primary liver sarcomas.[1-4]

The term undifferentiated embryonal sarcoma (HUES) was introduced by Stocker and Ishak in 1978 to describe a group of hepatic mesenchymal tumors that did not show evidence of differentiation. From 1978 to 2008 there was a record of 80 cases in the literature.[5,6] It is an aggressive mesenchymal tumor that occurs predominantly in the pediatric population between 6 and 10 years of age without gender predominance. In the adult population, it has a predisposition in the female gender.[7,8]

Early diagnosis is the key to increase the chances of long-term survival. Diagnosis is a challenge because it presents epidemiological, clinical, and radiological characteristics like other liver tumors and most of the diagnosis is by exclusion. It usually presents as a palpable abdominal mass sometimes accompanied by pain.

Computerized axial tomography is the ideal imaging study to characterize the lesion. A large, solitary lesion with a cystic component with well-differentiated borders with late enhancement is identified. Occasionally presents with a pseudocapsule and internal calcifications.

There is no consensus on the treatment of choice for these lesions la.[3] The treatment of choice is based on complete surgical resection, however, on some occasions with a poor prognosis if an R0 is not obtained.[2,3,11]

CASE REPORT

A 20-year-old female with current condition that began one month prior to admission with abdominal pain, located in the epigastrium radiating to the shoulder and back, intensity of up to 6/10, without extenuating or exacerbating factors. She received treatment for suspected acid-peptic disease without improvement. 48 hours prior to admission, the abdominal pain was exacerbated, accompanied by lipothymy, for which she was taken to this hospital. On her arrive she was found with tachycardia, generalized pallor, and a poorly defined, non-mobile, painful epigastric tumor was palpated approximately 5x5 cm, laboratory highlighted a hemoglobin of 7 g/dl, the rest without alterations. As part of its diagnostic approach, a simple and contrasted abdominopelvic tomography was performed (Figure 1- 3) with the presence of a hepatic lesion located in the left lobe of well-circumscribed ovoid morphology, discreetly lobulated margins of heterogeneous density and areas of hemorrhage, of 8.8x12.4x10.4 cm in its major axes, with a mass effect that displaced adjacent structures, discontinuation of its capsule to which the collection was associated, with extension to pelvic cavity. Due to the high suspicion of active bleeding at the level of the tumor, it was decided to perform interventional radiology embolization of the left hepatic artery (Figure 4 and 5). As part of the preoperative protocol, to assess the morphofunctional liver, a hepato-splenic scintigram was performed with SPECT/CT with evidence of a space occupying lesion in the non-functional left hepatic lobe and preserved hepatocellular function of the right hepatic lobe and segments III and IV (Figure 6). It was surgically approached with a Chevron-type wound. Hepatic bisegmentectomy (II and III) was performed with the findings of hemoperitoneum 500cc, liver tumor of 8x11x10 cm in segments II and III, rounded with lobulated edges with adhesions to the omentum, parietal peritoneum and stomach, macroscopically negative edges (Figure 7), with total bleeding of 600 cc, no trans-operative histopathological study was performed. The patient presented a favorable post-surgical evolution and was discharged. The histopathological report and immunohistochemistry conclude: High-grade sarcoma, compatible with HUES, Ki67 +, Vimentin + and CD56 + (Figure 8 and 9).

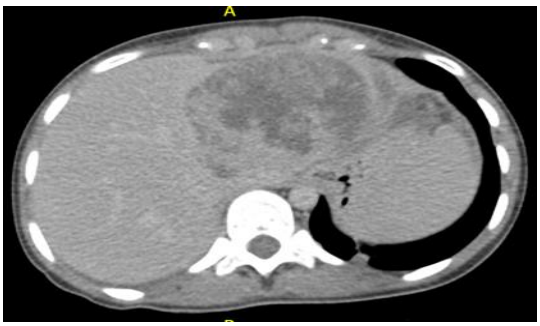

Figure 1: Simple phase: isodense and hypodense areas with respect to healthy liver parenchyma.

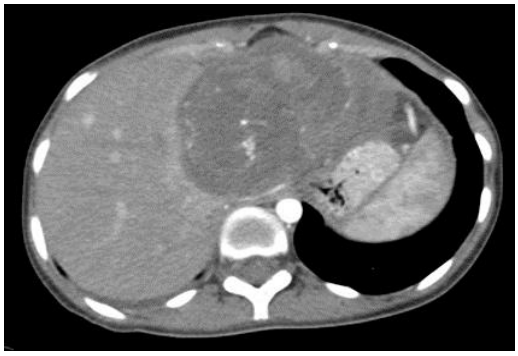

Figure 2: Arterial phase: peripheral and nodular enhancement similar to the aorta.

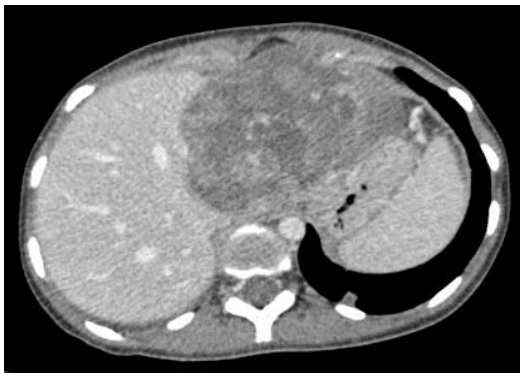

Figure 3: Portal phase: centripetal enhancement similar to the aorta.

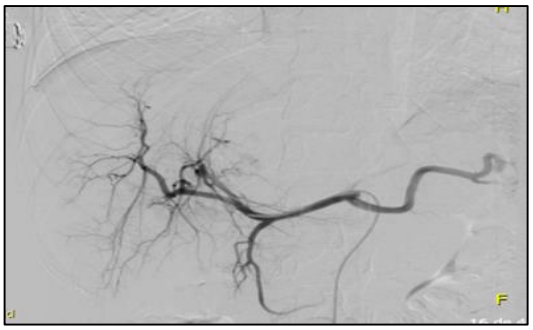

Figure 4: Adequate flow of contrast medium is observed towards both right and left hepatic arteries.

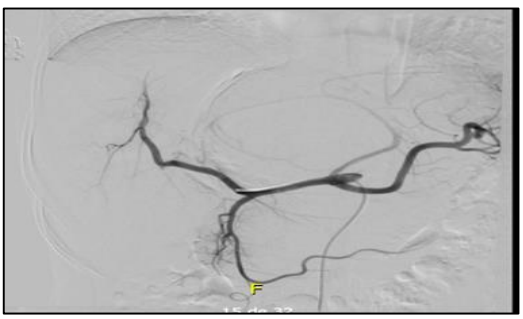

Figure 5: Absence of contrast medium flow towards the left hepatic artery due to embolization at its proximal level by the interventional radiology service.

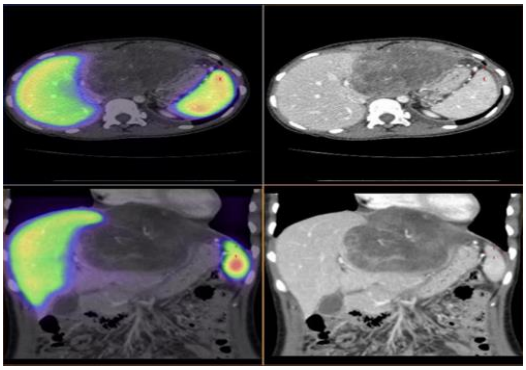

Figure 6: Abdominal SPECT fused with abdominal CT: morphofunctional correlation shows liver segments II and III nonfunctional.

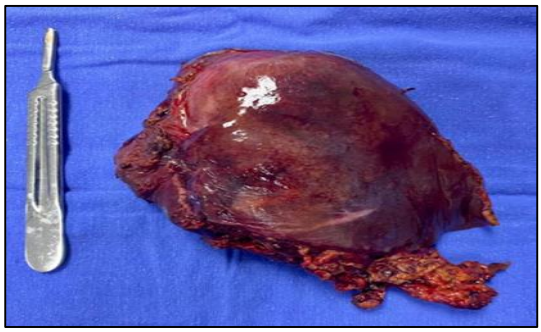

Figure 7: Surgical piece product of hepatic bisegmentectomy (II and III) on its anterior face.

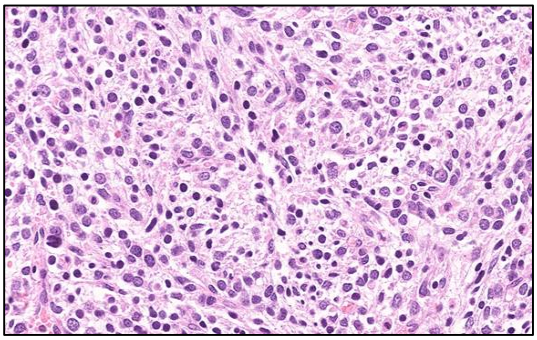

Figure 8: 40X HE polymorphic cells with poor cytoplasm and round to ovoid hyperchromatic nuclei or with granular chromatin and nucleolus.

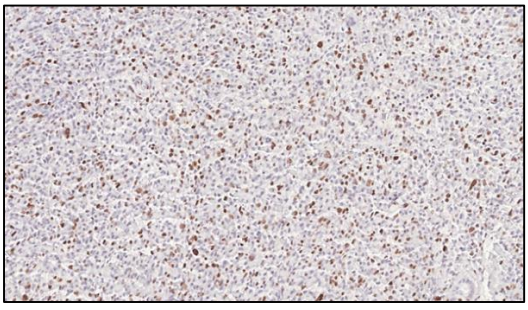

Figure 9: 25X Ki-67 positive in 55% of the nuclei of neoplastic cells.

DISCUSSION

Liver sarcoma has several histological lines including angiosarcoma (32%), leiomyosarcoma (29%), epithelial hemangioendothelioma (15%), undifferentiated embryonal sarcoma (7%), among others.[1-3] Its etiology is unknown, unlike of other hepatic neoplasms.[4]

Undifferentiated embryonal sarcoma (HUES) is an aggressive mesenchymal tumor that occurs predominantly in the pediatric population between 6 and 10 years of age without gender or ethnic predominance. It commonly involves the right lobe and usually has an asymptomatic course. It is the third most common primary malignant liver tumor after hepatoblastoma and hepatocellular carcinoma in this age group, corresponding to 9-15% of cases. In the adult population, it has a predisposition in the female gender.[7,8]

It usually presents as a palpable abdominal mass sometimes accompanied by pain. As secondary symptoms may occur fever, weight loss, anorexia, vomiting, diarrhea, lethargy, among others. Fever is usually associated with hemorrhage and necrosis. As the case of our patient, it can present with an acute abdomen with spontaneous rupture of the lesion in the abdominal cavity, related to rapid growth.[3,7] At the moment there is no tumor marker that increases suspicion or diagnosis, including alpha fetoprotein, a marker tumor that rises in most malignant hepatic tumors in HUES remains normal.[8]

A peculiar characteristic of this type of lesions and described by Moor et al as the "paradoxical appearance" is the architectural difference seen in ultrasonography and CT, since these lesions have predominantly solid characteristics on USG and a cystic appearance on CT.[8,9] It is generally a large (10-30 cm), solitary and well-circumscribed tumor with variable areas of hemorrhage, necrosis, and cystic degeneration. The solid component of HUES is sarcomatoid with a myxoid background. Cells are spindle-shaped or stellate with discrete nucleoli and poorly defined membrane. At the immunohistochemical level, most cases are positive for vimentin, desmin, alpha-1-antitrypsin, CD56, CD68, B-cell lymphoma and CD10.[5-7]

Spontaneous tumor-associated hepatic hemorrhage is a rare surgical emergency and accounts for 1% of referrals to liver clinics. The most common cause of spontaneous hepatic hemorrhage is hepatocellular carcinoma. Control of bleeding with preservation of as much liver function as possible is the primary goal of emergency management. There are few studies that recommend transcatheter arterial embolization (TAE) as the first-line method of acute hemostasis with subsequent definitive treatment.[10]

Surgical resection for primary liver sarcoma is considered a better treatment than chemotherapy, however dual therapy with surgery and chemotherapy is even better.[1]

The chemotherapy regimen used varies between institutions and oncologists since a standard treatment is not yet established, the regimens may include doxorubicin, ifosfamide, cyclophosphamide, vincristine and etoposide.[6,7] In pediatric population survival at 5 years is 86%.[6] Until 1990 a survival rate of 37% was reported, however, the prognosis has slowly improved with multimodal management that includes not only surgery but radiotherapy and chemotherapy, so survival has increase to 70-100% currently.[6,7]

Single photon emission computed tomography (SPECT)/CT with 99 m Tc sulfide colloidal generates images of the reticuloendothelial (Kupffer) cells of the liver. It has been shown to be an imaging biomarker that is related to liver function and can determine the functional tissue and non-functional before, during and after treatment. It is useful to complement the risk classification and promote the selection of targeted therapy in patients with liver tumors including surgery, intra-arterial embolization, radiotherapy and even evaluation of the liver transplant.[13]

PET-CT with 18 F-fluorodeoxyglucose is useful for staging the disease, detection of early recurrences and response to treatment after chemotherapy.[14]

The recurrence rate is higher during the first two years after surgery, and the risk is higher with positive surgical margins and in cases with spontaneous and/or iatrogenic rupture of the liver injury.[7] Poor prognostic factors are histological type, lymph node involvement, tumor rupture and R1/R2 resections.

CONCLUSION

In the literature there are few case reports of HUES since its description in 1978. It is a highly aggressive tumor of mesenchymal origin. Due to its ambiguous presentation, it represents a diagnostic challenge. It requires prompt and multidisciplinary management based on dual therapy of surgical resection and chemotherapy to improve the prognosis. As a result of its low incidence, there is no consensus or international guidelines on treatment, so more research is required to provide the appropriate tools for early diagnosis and offer the best management with the aim of reducing morbidity and mortality and increasing survival in patients.

In this article, we presented the case of a female patient with HUES as a cause of acute abdomen secondary to rupture of the tumor, that require initially interventional radiology embolization of the left hepatic artery and posteriorly surgical treatment.

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