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Enrique Moncada*, Vincenzo Vigorita, Alberto de San Ildefonso, Raquel Sánchez Santos

Unidad de Coloproctología, Servicio de Cirugía General y del Aparato Digestivo, Complejo Hospitalario Universitario de Vigo, Vigo, Spain

*Corresponding author.
E-mail address: emoniri@telefonica.net (E. Moncada).

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Calcifying nested stromal-epithelial tumor: A rare hepatic neoplasm☆

Tumor calcificante en nidos epitelial-estromal: neoplasia hepática excepcional

Calcifying nested stromal-epithelial tumors (CNSET) are rare primary liver tumors[1–6] that were first described by Ishak in 2001[1,3,4,6]. Only 43 cases have been published in the literature[2–9] (Table 1). These neoplasms present an apparently benign and indolent clinical course, attributable to their low malignant potential[10]. Surgical treatment with free margins usually provides high long-term survival rates.

We present a 21-year-old patient with no relevant history. Follow-up lab work showed an altered liver profile (gamma glutamyl transpeptidase [GGT] 122 IU/L). Physical examination revealed hepatomegaly. Abdominal ultrasound identified a hyper-isoechoic focal lesion, apparently calcified, with a lobulated shape and limited vascularization. Magnetic resonance imaging (MRI) and computed tomography (CT) scan (Fig. 1A–B) showed a large, irregular, heterogeneous mass with multiple calcifications, with irregular enhancement in the arterial phase and washout in the portal phase. The initial diagnosis was fibrolamellar hepatocellular carcinoma (FHCC) in segments IV, V, VI, VII and VIII. Tumor marker levels (Alpha-fetoprotein, carcinoembryonic antigen and CA 19.9) were normal. Total liver volume was 1172.83 cc. The calculated residual volume percentages were: 42.2% for segments II-III (495.59 cc) and 48% for segments I-II-III (562 cc). Due to the suspicion of FHCC, surgery was indicated, and we performed right trisectionectomy by laparotomy as well as lymphadenectomy of the hepatic hilum and celiac trunk. The postoperative period transpired without incident, and the patient was discharged on the 7th postoperative day.

The pathological study reported a whitish nodular tumor measuring 21 × 13 × 8 cm with multiple calcifications, areas of ossification and lymphadenopathies without neoplasm. Immunohistochemistry showed the cells were positive for Vimentin, Actin, WT-1, CD56, CD99, CD117 with Ki67:1% (Fig. 1C–D). These results were compatible with CNSET with free surgical margin, no perineural invasion or lymphovascular permeation.

The case was presented to the Oncology Committee, who decided to monitor the patient in an outpatient clinic without adjuvant chemotherapy because there is no clear benefit to justify adjuvant therapy. Six months after surgery, the patient remains asymptomatic and disease free. Follow-up studies included laboratory testing with liver panel and CT scan.

CNSET have been described in the literature with various terminologies: ossifying malignant mixed epithelial and stromal tumor, ossifying stromal epithelial tumor, and desmoplastic nested spindle-cell tumor of the liver[1]. It was Markhouf who proposed the term CNSET because it incorporates all the characteristics of the tumor.

Despite their exceptional nature, we know that they present more frequently in young (≤20 years) or pediatric patients (77%) and females (70%) (Table 1). They are usually located in the right hepatic lobe (64%), and in 40% of cases their diagnosis is incidental (Table 1). The association of CNSET with hormonal alterations is notable (36%): Cushing syndrome or cushingoid symptoms (25%)[5,7], Klinefelter syndrome[4] and oral contraceptives. Some authors have tried to link the development of these tumors with hormonal alterations, but this has not been proven. In four patients, it was associated with Beckwith-Wiedemann syndrome (overgrowth syndrome with increased risk of developing cancer)[2,8]. Our case did not present an association with these pathologies.

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Table 1 – Clinical and pathlogical características of 44 cases of calcifying nested stromal-epithelial tumor.

Journal	Age	Sex	Síntoms	Surgical treatment	Chemotherapy	Follow-up
Heywook, Cancer 2002	28	Female	Incidental	Right trisegmentectomy	No	Recurrence 72 months → RF
Hill, Am J Surg Pathol 2005	2	Male	Incidental	Right partial hepatectomy	post-op CTx	Living, 84 months
	6	Female	Incidental	Liver resection in right lobe	No	Living, 58 months
	6	Female	Incidental	Liver resection in right lobe	No	Living, 8 months
	14	Female	Incidental	Liver resection in right lobe	post-op CTx	Recurrence 11 years
Herema Mc Kenney, Am J Surg Pathol 2005	2	Male	Incidental	Hepatic excision	No	Living, 8 months
	3	Female	Incidental	Liver resection	Pre-op and post-op CTx	Living, 6 months
	4	Male	Incidental	Enucleation	post-op CTx	Living, 36 months
	11	Female	Characteristics of Cushing + abdominal mass	Left lobectomy	No	Living, 24 months
	12	Female	Características of Cushing + abdominal mass	Right hepatectomy	No	Living, 168 months
	14	Female	Íleo	Liver resection	post-op CTx	Recurrence 12 months (Local) →CTx
Brodsky, Pediatr Dev Pathol 2008	17,5	Female	Características of Cushing + abdominal pain	Left lobectomy + partia hepatectomy	No	Recurrence 12 months (local and lymph nodes) → liver trasplant
Wirojanan, J Pediatr Hematol Oncol 2008	2	Female	Not determined	Liver resection	post-op CTx	Living, 84 months
Meir, Pediatr Dev Pathol 2009	2,5	Female	Incidental	Lobectomía derecha	No	Living, 8 months
Makhlouf, Am J Surg Pathol 2009	2	Female	Incidental	Right partial hepatectomy	No	Living, 6 months
	14	Female	Incidental	Right partial hepatectomy	post-op CTx	Living, 264 months
	15	Female	Incidental	Right partial hepatectomy	No	Living, 151 months
	16	Male	Características Cushing	Right partial hepatectomy	No	Living, 56 months
	18	Female	Incidental	Liver transplant	No	Éxitus 50 months→ complicaciones postoperatorias. No Recurrence
	19	Male	Incidental	Needle biopsy (right lobe lesion)	post-op CTx (after recurrence)	Local recurrence 168 months→ RF ablation and CTx
	32	Female	Incidental	Right partial hepatectomy	No	Living, 13 months
	33	Female	Incidental	Right partial hepatectomy	No	Living, 14 months
Rod, Eur J Endocrinol 2009	17	Female	Características of Cushing + abdominal mass	Left resection	No	Living, 30 months
Grazi, Pathol Res Pract 2010	25	Female	Diarrhea + recurrent abdominal pain	Right lobectomy extended to caudate and segment IV	No	Living, 6 months
Oviedo Ramírez, Ann Diagn Pathol 2010	33	Male	Nonsoecific abdominal pain + dyspepsia	Left lobectomy	No	Living, 15 months
Marin, Am Surg 2010	33	Male	Epigastralgia + pirosis + regurgitación	Unknown	Unknown	Unknown
Wang, Int J Surg Pathol 2011	34	Female	Incidental	Left lobectomy	No	Living, 42 months

Table 1 (*Continued*)

Journal	Age	Sex	Síntoms	Surgical treatment	Chemotherapy	Follow-up
Homman, J Gastrointest Cancer 2011	16	Female	Incidental	Liver transplant	Post-op CTx	Recurrence after 28 months with lung metástases Éxitus, 37 months
Assmann, Hum Pathol 2012	16	Male	Palpable abdominal mass	Liver transplant (right/left liver lobe lesion)	Pre-op CTx	Living, 24 months
	3	Female	Constipation	Partial left hepatectomy	Post-op CTx	Living, 5 years
Geramizadeh, Indian J Pathol Microbiol 2012	8	Male	Características of Cushing + weight gain + abdominal pain	Extended right hepatectomy	No	Éxitus, 10 days (due to postoperative complications)
Ghodke, J Postgrad Med, 2012	9	Male	Abdominal pain + fever + jaundice + weight loss	Liver segmentectomy	Not determined	Living, 12 months
Malowany, Pediatr Dev Pathol 2013	2	Female	Incidental	Right resection	Not determined	No Recurrence
Procopio, World J Hepatol 2014	23	Female	Abdominal distension + dispepsia	Extended left hepatectomy	No	Living, 21 months
Samarghandi, Clin Nucl Med 2015	11	Female	Weight gain + increased apetito + abdominal pain	Right lobe	Not determined	Not determined
Schaffer et al.[6], Clin Imaging 2016	14	Female	Abdominal distension + hinchazón mejillas Síndrome Cushing	Liver transplant (left liver lobe lesion)	Pre-op CTx	Living, 10 months
Weeda et al.[5], J Med Case Rep 2016	16	Male	Características Cushing + weight gain + abdominal distension	Right and left trisegmentectomy	No	Living, 13 years
Khoshnam et al.[2], Eur J Med Genet 2017	14	Female	Características Cushing+ Hinchazón y dolor abdominal	Liver transplant (Lesión lóbulo hepático derecho)	Pre-op CTx	Living,
Tehseen et al.[7], Pediatr Transplant 2017	13	Female	Dolor abdominal + abdominal distension Cushing syndrome	Liver transplant (Lesión lóbulo hepático derecho)	Pre-op CTx	Living, 28 months
Garg et al.[8], Indian J Nucl Med 2017	8	Female	Pancitopenia	Liver resection; liver transplant	Post-op CTx in recurrence	2 months lymoh node and bone recurrence →CTx + Bone RT
Meletani et al.[9], World J Gastroenterol 2017	31	Male	Abdominal pain + weight loss	Right triseccionectomy + caudate resection + RF segment II	post-op CTx	Local recurrence after 6 months + p presacral nodule → chemoembolización + CTx after (×4) Éxitus: 15 months after surgery
Tsuruta et al.[4], World J Surg Oncol 2018	20	Male	Incidental	Right hepatectomy	post-op CTx	Éxitus: 2 months due to local recurrence and metástases in extrahepátic lymph nodes→ chemoembolización and RF
Olin et al.[3], Int J Surg Case Rep 2020	15	Female	Dolor abdominal	Right trisegmentectomy	No	Living, 12 months, sin Recurrence
Pérez, 2020	21	Female	Asintomátic	Right triseccionectomy	No	Living, 6 months, sin Recurrence

Post-op: postoperative; Pre-op: preoerative; CTx: chemotherapy; RF: radiofrecuency.

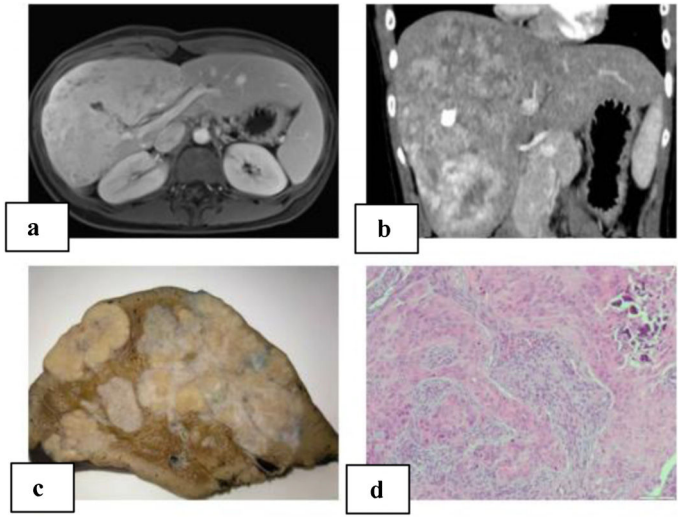

Fig. 1 – A) MRI of the liver: large, heterogeneous, polylobulated mass measuring 11.5 × 10 × 19 cm in segments IV, V, VI, VII and VIII with hypervascular uptake, with no clear washout in the portal and late phases, maintaining foci with no uptake at the central level, which coincided with the calcification on the CT scan; diffusion was slightly restricted; characteristics were compatible with fibrolamellar carcinoma; B) abdominal computed tomography: a large heterogeneous hepatic mass with irregular contour and heterogeneous density occupied segments IV, V, VI, VII and VIII, with multiple calcifications; irregular enhancement in arterial phase, and contrast washout in portal phase; C) macroscopic image: hepatectomy specimen measuring 24 × 17 × 19 cm and weighing 1500 g. It is almost entirely occupied by a 21 × 13 × 8 cm nodular whitish tumor that is in contact with the hepatic capsule and is away from the parenchymal resection margin; D) microscopic image with hematoxylin-eosin (10× magnification): proliferation of epithelioid and spindle-shaped cells with formation of osteoid; nests are observed with a central epithelial appearance, surrounded by spindle-shaped cells; calcified area.

In terms of diagnosis, the lab analysis highlights normal liver function as well as the usual tumor markers[1,3], although elevated GGT has been described in some cases[5], including ours. On CT and MRI, it is characteristic to observe a large, well circumscribed, heterogeneous mass with dense calcification and macrolobulated margin[1,3,4,6]. The most common radiological differential diagnosis is hepatoblastoma, followed by FHCC. Confirmation of the diagnosis by preoperative biopsy has been described[3]. Geramizadeh published low profitability in a cohort of 12 patients with CNSET, where the diagnosis was reached in only three cases (25%). At our hospital, the use of liver biopsy is unusual and only reserved for uncertain diagnoses.

The standard treatment of CNSET is surgical resection, obtaining free margins[1,3–5]. The technique used has varied from partial resections to liver transplantation, which occurred in eight cases, depending on the extension and location of the tumor (Table 1).

Chemotherapy (CTx) was used in 38% of the cases of the series analyzed. Five cases received preoperative CTx, showing no effects of reduction or necrosis in the imaging tests or in the surgical specimens. Twelve patients received postoperative CTx, 50% as treatment for postoperative recurrence (Table 1). The regimens administered were similar to those for sarcoma or hepatoblastoma[4–6]. Today, its role in the prevention or treatment of recurrence is unclear[1,4,5]. Three were treated with radiofrequency (RF)[4], and only Makhlouf describes a successful result with this therapy.

80% (35/44) of the patients did not experience disease recurrence. This occurred in nine patients with an interval from two months to 11 years[4,8,9]. Most recurrences were local, but they were found in several locations: lymph nodes, lungs or bone (Table 1). The follow-up described in the literature varies from two months to 22 years. In total, 7% of patients died from the disease, and 14% are living with recurrence (Table 1).

The prognosis is uncertain, but growth is usually slow, with low malignant potential[1,4,5]. Most cases have long-term survival after resection[4] and often remain recurrence-free if the resection margins are negative[3]. A close and sustained follow-up is necessary, with lab work and imaging studies due to the risk of long-term recurrence[2–5].

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María Pérez Reyes*, Belinda Sánchez Pérez, Francisco Javier León Díaz, Julio Santoyo Villalba, Julio Santoyo Santoyo

Hospital Regional Universitario, Málaga, Spain

*Corresponding author.
E-mail address: maria.perezreyes1991@gmail.com (M. Pérez Reyes).

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Covid no-related surgical emergencies during Covid-19 time. Case report: broad ligament internal hernia with associated small bowel necrosis☆

Urgencias quirúrgicas no relacionadas con la pandemia COVID-19. Caso clínico: hernia interna yeyunal complicada con necrosis intestinal en el ligamento ancho útero-ovárico

Internal hernia is responsible of less than 1% of intestinal obstruction. Hernia through the broad ligament is a rare condition reported as less than 7% of all internal hernias.[1] During Covid-19 pandemia patients are scared about going to the hospital with an high risk of getting infected and they tend to underestimate the severity of their symptoms. Consequently they arrive to the hospital with a late-stage acute pathology. This state of fact is illustrated by this case report.

The endpoint of the case report is to illustrate a case of a complicated bowel obstruction in which the intestinal resection could have been avoided or minimized if the patient had gone earlier to the emergency.

In the context of the SARS-Cov-2 pandemic, we have detected in emergency services more complicated cases of acute pathologies. We have especially detected cases of periappendicular abscesses and cases of Hinchey 3 and 4 diverticulitis that in our center we usually find in an earlier stade.

The case concerns a 43 years old female with medical history of laparoscopic resection of uterine cyst 15 years ago (no information about the exact position). The patient came to the hospital 24 hours after symptoms starting. She had abdominal pain, nausea and vomiting since the day before. Beacause of the fear of getting the SARS-CoV-2 infection she decided not to come to the hospital despite the symptoms waiting 24 hours.

At the emergency admission the patient presented abdominal distension and guarding at low abdominal quadrants. Nausea and vomiting were still present. Blood test showed an inflammatory syndrome with leukocytosis (12460 G/l of white cells), normal C-reactive protein, LDH at 232 U/l and light anemia (10,9 g/dl of haemoglobin). The CT-Scan showed a

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