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Unicentric Castelman Disease: A Rare Indication for Major Hepatectomy

Horea Bocse[1], Caius Breazu[2]*, Cristina Pojoga[3]**, Ioana Iancu[4]**, Sandu Branzilă[4], Adrian Bartoș[5]

***Corresponding author:**
Department of Anesthesia
City County Hospital, Cluj-Napoca
Iuliu Hatieganu University of Medicine and Pharmacy, Cluj-Napoca, Romania
E-mail: csbreazu@yahoo.com

**Same contribution as the main authors

[1]Department of Surgery, Prof. Dr. Octavian Fodor Regional Institute of Gastroenterology-Hepatology, Cluj-Napoca, Romania
[2]Department of Anesthesia, City County Hospital, Cluj-Napoca
Iuliu Hatieganu University of Medicine and Pharmacy, Cluj-Napoca, Romania
[3]Department of Gastroenterology, Prof. Dr. Octavian Fodor Regional Institute of Gastroenterology-Hepatology, Cluj-Napoca, Romania
UBB Med, Babes Bolyai University, Cluj-Napoca, Romania
[4]Department of Surgery, Medicover Hospital, Suceagu, Cluj, Romania
[5]Department of Surgery, Prof. Dr. Octavian Fodor Regional Institute of Gastroenterology-Hepatology, Cluj-Napoca, Romania
Iuliu Hatieganu University of Medicine and Pharmacy, Surgical Department, Cluj-Napoca Romania
Medicover Hospital, Surgical Department, Suceagu, Cluj, Romania

ABSTRACT

This report presents a rare case of Unicentric Castleman Disease (UCD) necessitating a major hepatectomy in a 53-year-old Caucasian male with an anomalous hepatic mass. Despite the non-specific findings on MRI and CT scans, the use of intraoperative ultrasonography revealed critical tumor proximity to the left hepatic artery, guiding the decision for a left hepatectomy. Post-surgical histopathological analyses confirmed the UCD diagnosis, emphasizing its rarity in hepatic contexts. The case underlines the importance of combined imaging modalities, especially intraoperative assessments, and thorough histopathological evaluation in diagnosing atypical liver tumors, highlighting the rare presence of Castleman disease in liver.

Key words: Unicentric Castleman Disease (UCD), hepatic tumor, intraoperative ultrasonography

INTRODUCTION

Castleman disease (CD), often referred to as angiofollicular lymph node hyperplasia, is an uncommon lymphoproliferative disorder that predominantly manifests in the lymph nodes (1). This group of lymphoproliferative disorders share common histopathological features (2). Unicentric CD (UCD) involves one or more enlarged lymph nodes in a single region of the body that present specific histopathologic characteristics (3,4). These features can be categorized into three main subtypes: hyaline vascular (HV), plasma cell (PC) and mixed (1-4). The etiology of the disease is poorly understood but several theories have been proposed: Lymphoid Hyperplasia, Viral Infections (unlike multicentric CD-MCD, UCD is not associated with human herpesvirus 8 HHV-8 or HIV infection),

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Immune dysregulation, Cyotokine Production (Interleukin-6 might play a role in the pathogenesis), Cytogenetic abnormalities (modifications in chromosome segment 12q13-15)(5).

UCD symptoms are usually related to mass pressing against adjacent structures and sometimes (more specific to MCD) patients present with systemic symptoms such as: fever, night sweats, weight loss or fatigue. Laboratory studies are usually normal in these patients (6). Accurate diagnosis typically involves a combination of imaging studies (CT, MRI, PET-CT), blood tests and most definitively a biopsy of the involved lymph node (4-6). The annual incidence of CD in the US ranges from 6500 to 7700 of which 75% are estimated to be UCD (7). The long-term disease-free survival in patients with resectable tumors is high (4% mortality over 10-years of follow-up) (8,9,20).

Despite a plethora of individual case reports and studies, cohesive understanding has been somewhat elusive. This case report highlights the intricacies of CD diagnosis, the rarity of hepatic involvement and the importance of individualized patient care.

CASE REPORT

A 53-year-old Caucasian man, devoid of significant medical history and presenting no specific symptoms, approached our unit with a recently diagnosed tumor mass in the left liver lobe.

Liver MRI, enhanced with Primovist, depicted a tumor mass in segment III, measuring 32 x 28 mm. It exhibited a low signal on T1 and a high signal on T2, coupled with arterial enhancement and relative washout during the venous phase (*fig. 1*). The CT scan identified enlarged lymph nodes at the hepatic pedicle, but no extra-hepatic tumor masses were observed. Laboratory assessments were unremarkable, showing normal liver enzymes and tumor markers, with alpha-fetoprotein (AFP) and Carcinoembryonic Antigen (CEA) within standard limits. Pre-surgical liver elastography (Fibroscan) did not display any pathological alterations, registering as F0 Metavir. Since we had no contraindication for radical treatment, corroborated with the imagistic information, the indication of surgery was made.

During surgery, the tumor mass was observed on the diaphragmatic surface of the left lobe. A routine intraoperative ultrasound (IOUS) was conducted, revealing the tumor's proximity to the left hepatic artery. Given this arterial involvement, a left hepatectomy was considered appropriate, together with a hepatic pedicle lymphadenectomy the standard technique of our team when treating hepatic tumors, for extemporaneus histopatological exam, which showed no malignant cells (*figs. 2* and *3*).

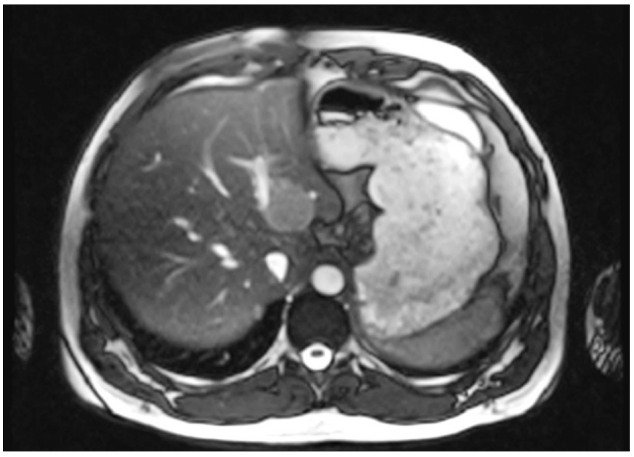

Figure 1 - MRI showing a T2 hyperintensity liver tumor mass in the left lobe

Postoperatively, the patient underwent 24-hour monitoring in the Intensive Care Unit (ICU). The recovery process was uncomplicated, marked by early oral feeding on postoperative day 1, bowel movement normalization on day 2, and drain removal by day 4. An abdominal ultrasound performed on day 3 post-op did not flag any notable alterations. The patient received discharge on postoperative day 5.

Histopathological evaluation of the surgical specimen confirmed the diagnosis as hepatic UCD, of the hyaline vascular subtype (*fig. 4 a, b*), without extrahepatic (hepatic pedicle) lymph nodes involvement.

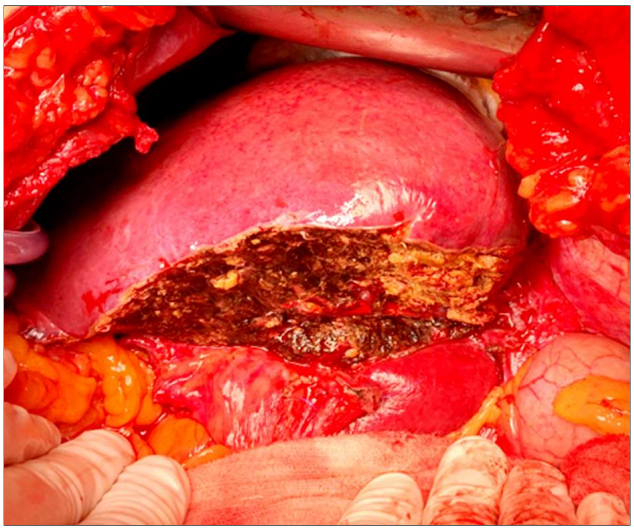

Figure 2 - Liver remnant tissue after left hepatectomy (from the personal library of dr Bartos A.)

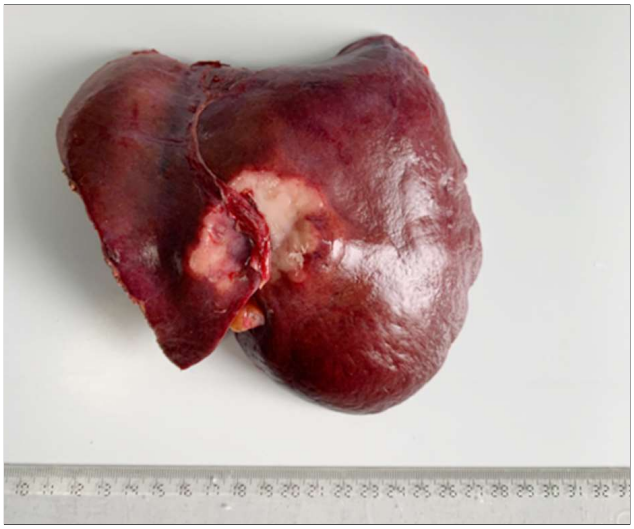

Figure 3 - Surgical specimen showing the liver mass on the diaphragmatic surface (from the personal library of dr Bartos A.)

Periodic assessment revealed no evidence of recurrence (3 years now of writing the article).

DISCUSSIONS

This case delineates the diagnostic challenges posed by an anomalous hepatic mass in a 53-year-old Caucasian male, emphasizing the nuanced intricacies of hepatological investigations.

The MRI, while insightful with its display of enhancement patterns, was not conclusively diagnostic on its own. This reinforces the idea that when faced with ambiguous liver tumors, a combination of both MRI and CT scans can provide a clearer picture (11,12) Although the CT scan identified enlarged lymph nodes suggesting possible metastatic involvement, the absence of extra-hepatic tumors and consistent laboratory findings presented a diagnostic challenge.

This situation underscores the notion that imaging and blood tests, while useful, are not always definitive when deciphering liver tumor etiologies, the surgical treatment being the only treatment available for a hepatic mass with imagistic suspicions of malignancy, a biopsy being not mandatory. The pivotal role of intraoperative ultrasonography (IOUS) emerged during the surgical intervention. Its utility in discerning the proximity of the tumor to the left hepatic artery was instrumental in steering the surgical strategy towards a left hepatectomy. This emphasizes the role of intraoperative imaging modalities in formulating and refining surgical tactics (13).

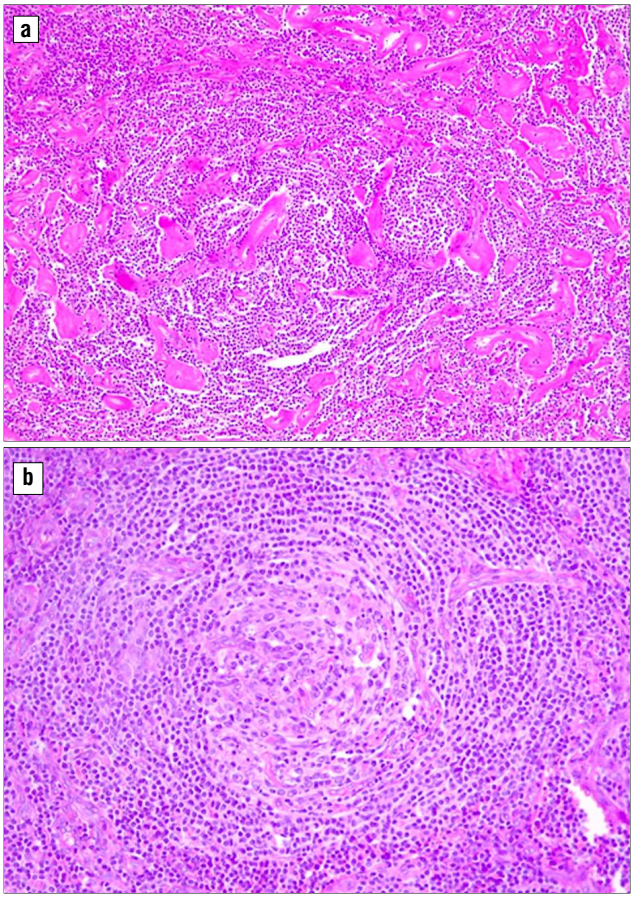

Figure 4 - Histopathological findings.
(a) Hematoxylin and eosin stain, 10x: Hyalinized blood vessels appearing to penetrate an aggregate of germinal center cells.
(b) Hematoxylin and eosin stain, 20x: Germinal centers with hyaline deposits.

Pathologically, the diagnosis of UCD, specifically the hyaline vascular subtype, further amplifies the complexity of this case. The rarity of this pathology in hepatic presentations underlines the paramount importance of histopathological analyses, especially when confronted with diagnostic ambiguities (14,15).

CONCLUSIONS

Advanced imaging techniques were crucial for detailed tumor assessment and surgical planning. The presence of the tumor, guided by imagistic suspicion was sufficient for surgical indication. IOUS effectively guided the decision for left hepatectomy. UCD, particularly the hyaline vascular subtype, poses a unique diagnostic challenge in hepatic presentations, which is rare, but is highlighted by our case. Histopathological evaluation remains vital for diagnosing rare entities like UCD.

Conflict of interest

None.

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Ethical statement

We had all consents signed by the patient.

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