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Case Report

Castleman's disease: Laparoscopic resection of unicentric disease in the hepatic hilum: A case report

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ABSTRACT

Introduction and importance: Castleman's disease was first reported by Benjamin Castleman et al., in 1954 and described it as a sporadic lymphoproliferative disorder. The pathophysiology to this day is still unknown, although IL-6 is suspected to play an important role. Preoperative diagnosis is challenging due to its non-specific symptoms, and that imaging cannot clearly distinguish the disease from other processes. High clinical awareness is necessary to reach a diagnosis. If the disease is localized, complete recovery can be achieved through surgery.
Case presentation: Patient is a 68-year-old woman with a three-month history of recurrent episodes of fever, myalgias, and night sweats. She started to experience lower abdominal pain and presented to the emergency room. A contrast-enhanced abdominal computed tomography revealed a 5 cm well-circumscribed focal heterogeneously enhancing hyperplastic mass between the portal vein and the inferior vena cava. After successful laparoscopic surgery, the mass was resected, and the patient fully recovered. Unicentric Castleman's disease was the final diagnosis.
Discussion and conclusion: Castleman's disease is an uncommon pathology with a challenging diagnosis. When approaching an abdominal mass, unicentric Castleman's disease should always be a differential diagnosis, as treatment can be curative with surgical resection. With the advent of laparoscopic and robotic surgery, these techniques can improve patients' outcomes in these rare pathologies, especially when they appear in complex regions.

Introduction

Castleman's disease is a rare lymphoproliferative disorder (Incidence of 0.001–0.05%) of unknown etiology [1,2]. It's a benign tumorous process of the lymphocytes whose multiplications lead to an excessive enlargement of the lymph nodes [2,3]. It can be classified based on the involvement of one to several lymph nodes [3]. It mainly affects chest lymph nodes, yet, on rare occasions, the mesenteric lymph nodes can be affected. Surgery is the only way to ensure full recovery, and a laparoscopic approach can improve patients' recovery and results [3,4].

We present the case of a 68 year old woman with abdominal pain, a mass was identified close to the portal vein and after successful surgery she recovered completely. Unicentric Castleman's disease was the final diagnosis.

This manuscript adheres to SCARE guidelines [16]

Case Presentation

Patient is an otherwise healthy 68-year-old woman with a past medical history of cholecystectomy and hysterectomy. She had a three-month history of recurrent episodes of fever, myalgias, and night sweats. One month prior to admission, she started to experience lower abdominal pain. At first, the pain was mild and appeared soon after food intake, yet, as time passed, the pain migrated to her upper abdomen, became constant, and didn't mitigate even after analgesics; thus, she presented to the emergency room.

On clinical examination, a tachycardic patient with abdominal pain was encountered. She had moderate pain on touch on her upper

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abdomen without tenderness. No masses or lymph nodes were discovered at that time. A basic laboratory workup was made, and the results were within the normal range; then, an abdominal ultrasound was completed, but the results were inconclusive. Thus, a contrast-enhanced abdominal computed tomography was requested. It revealed a 5 cm well-circumscribed focal heterogeneously enhancing hyperplastic mass resembling a lymph node between the portal vein and the inferior vena cava; the mass had a capsule and did not invade any organs. No lymph nodes or other masses were detected (Fig. 1A and B).

Neuroendocrine tumors, lymphomas, or paragangliomas were among the differential, and due to the high risk of bleeding and spread of the tumor, a biopsy was ruled out. Therefore, surgery was decided with a laparoscopic approach. We were assisted by an expert laparoscopic surgeon with broad experience in liver and biliary surgery.

At surgery, the lesser sac was opened to reach the hepatic hilum. The lower part of the caudate segment of the liver was tractioned, and the 5cm mass was exposed. Using blunt dissection, polymer ligating clips, and bipolar energy (Ligasure, Medtronic, Dublin, Ireland), the white, soft, and vascularized mass was released entirely from the inferior vena cava (Fig. 2A and B).

The rest of the procedure was completed without complications. The patient's postoperative course was uneventful, liquid diet was started on the first postoperative day, and she was discharged on her third postoperative day without complications.

Pathology revealed a 5 × 3 cm soft mass; it had a whitish fibrous capsule surrounded by vascular tissue. On microscopic evaluation, an enlarged lymph node with multiple lymphoid follicular and fibroblastic proliferation was identified. It had numerous fibrotic septa with hyalinised vessels. Immunohistochemistry study showed positivity for CD21, CD20, and CD23. Unicentric Castleman disease with a hyaline-vascular subtype was the final diagnosis (Fig. 3A and B).

The patient is doing well on follow-ups. We do annual checkups that include an abdominal CT, complete blood count, and IL6. She is completely asymptomatic without signs of recurrence one year after surgery.

Discussion

First reported by Benjamin Castleman et al., in 1956 [1]. Castleman's disease (CD) is a rare non-clonal lymphoproliferative disease [2,3]. Although it was discovered over five decades ago, its pathophysiological basis is still under study [3]. It is believed that this disease occurs due to a deregulated interplay among inflammatory mediators, particularly IL-6, in response to human herpesvirus 8 (HHV-8), viral, neoplastic, or inflammatory pathways [2,3]. The affected cells secrete IL-6, which induces vascular endothelial growth factor production, further enhancing vascular proliferation inside the lymph node and IL-6 production of endothelial cells [3,4]. This excess results in lymphoproliferative proliferation, excessive expansion of lymph nodes, and systemic manifestations [3]. Our patient was HHV-8 negative and had no history of neoplastic disease. CD encompasses two distinct clinicopathological disorders, unicentric Castleman's disease (UCD), in which only one anatomic lymph node is affected, and multicentric CD, characterized by generalized lymphadenopathy with an aggressive clinical course with malignant potential [2,3]. Histopathologically, there are three primary subtypes; hyaline-vascular (70–80%), plasma cell, and mixed [4]. The hyaline vascular type is the most common and is characterized by follicular hyperplasia and its germinal centers exhibiting capillaries with hyaline matter [3,5]. All three variants present can present clinically with lymphadenopathy or systemic symptoms [3,6].

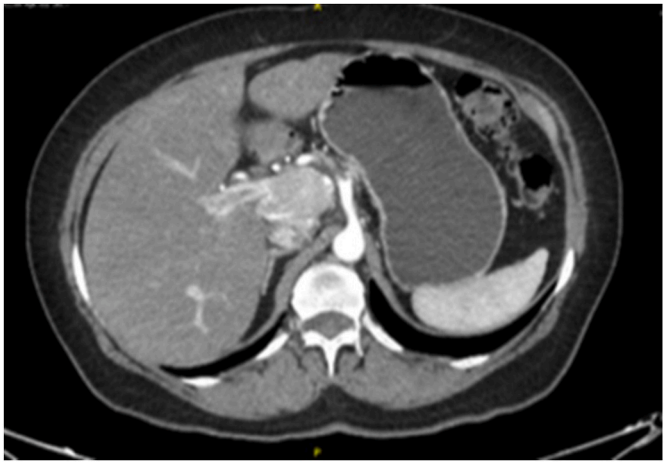

Fig. 1A. CT, revealing a lymph node between the portal vein and the inferior vena cava.

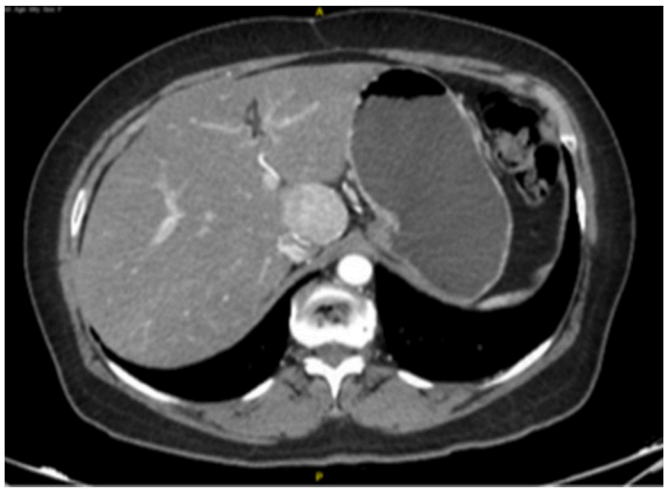

Fig. 1B. CT, revealing the heterogeneously enhancing hyperplastic mass.

UCD is a rare disease with 21 cases per million; it seems to have a slight predisposition with the female population between the third or fourth decades of life [2,3,7]. It usually affects the lymphoid tissues of the thorax (70%), neck (15%), abdomen-pelvis (12%), and axilla (3%) [5]. Mesenteric UCD is rare, with less than 100 cases reported worldwide [2,3]. Our patient had a UCD with a hyaline-vascular subtype detected in her abdomen. UCD is usually discovered incidentally during imaging studies performed for other reasons [2,7]. As the lymph node grows, the patient may present with symptoms related to the compression of nearby organs. (Dyspnea, cough in the chest, whereas vomiting or abdominal pain when it's located in the abdomen or retroperitoneum.) [3,8] As our patient experienced.

As symptoms are vague and non-specific, preoperative diagnosis without imaging studies is extremely difficult; therefore, high clinical awareness is critical [3,4,9]. Imaging studies such as CT, positron emission tomography, magnetic resonance imaging, and echography can reveal the location and characteristics of the lymph mass, yet, many tumors including GIST, neurogenic tumors, or ectopic pheochromocytoma, can show similar radiological features making the differential even more challenging [4,5,10].

An abdominal CT of UCD usually shows a homogeneous solitary mass with contrast enhancement without nodes or satellite nodules [10,11]. As it was found on our patient.

Laboratory evaluation can show positivity for Human Immunodeficiency Virus or HHV- 8 [3,12]. It can also indicate elevated plasmatic levels of cytokines such as IL-6 and IL−10; nonetheless, they are not routinely requested in clinical practice [4,7]. Final diagnosis is made by pathology; a biopsy by fine-needle aspiration can be helpful; nonetheless, achieving an adequate amount of tissue, the possibility of spreading tumor cells, and the risk of severe bleeding in hypervascular mass should

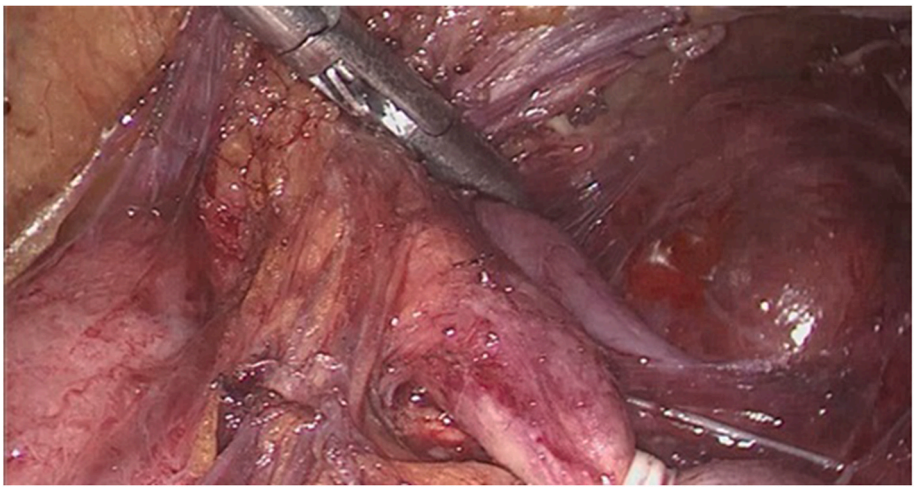

Fig. 2A. Laparoscopy, the mass is seen attached to the inferior vena cava.

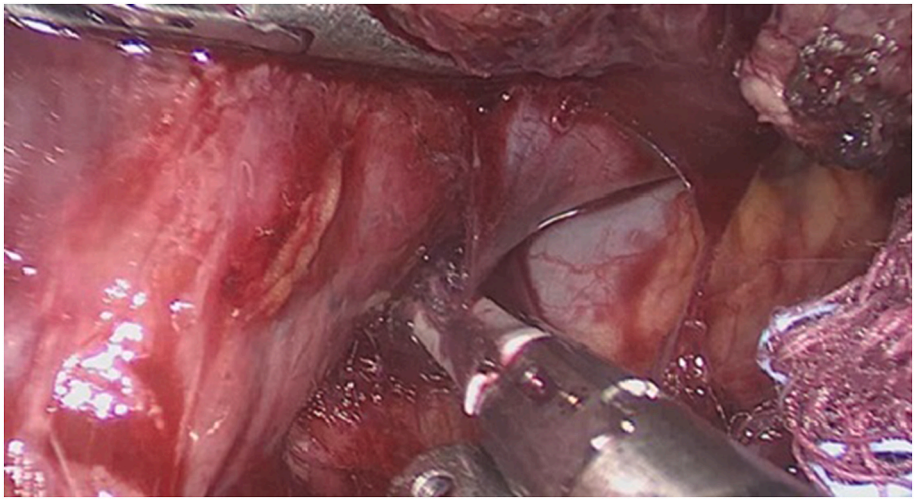

Fig. 2B. Laparoscopy, the mass is being released with blunt maneuvers.

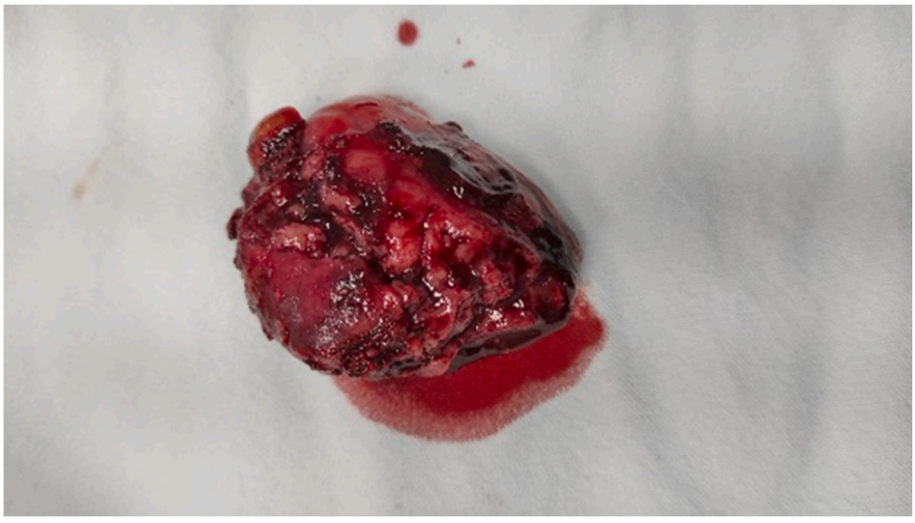

Fig. 3A. 5 × 3 cm soft mass with a whitish fibrous capsule surrounded by vascular tissue.

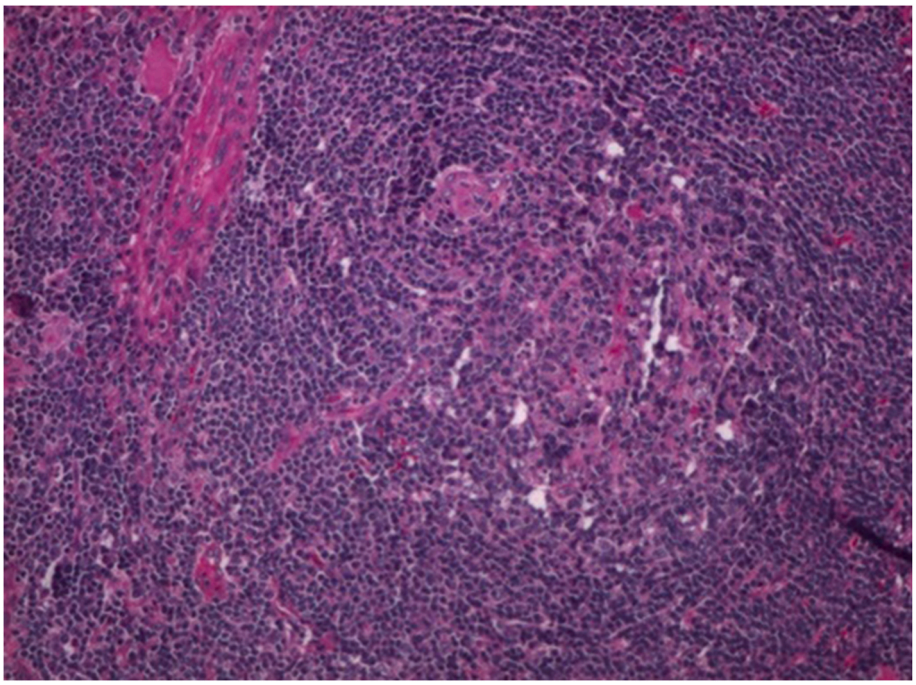

Fig. 3B. Enlarged lymph node with multiple lymphoid follicular and fibroblastic proliferation, numerous fibrotic septa with hyalinised vessels.

always be considered [4,13]. Even though UCD is not a malignant condition but many dangerous tumors, including Non-Hodgkin lymphoma, paraganglioma, leiomyoma, leiomyosarcoma, fibrosarcoma, and amyloidosis, have been associated with it [3,5]. The differential must also include diseases with lymphadenopathy, including tuberculosis, lymphadenitis, abscess, sarcoidosis, HIV, toxoplasmosis, among others [1,3,6]. In our patient, as the mass was discovered in an area of major vascular structures and had contrast enhancement, surgery was decided instead of a biopsy to prevent complications.

Complete surgical resection is the only curative approach. Patients with UCD who underwent surgery had higher overall survival (95.3%), 3-year disease-free survival (89.7%), and 5-year disease-free survival (81.2%) [10,14].Since abdominal UCD is a rare disease, most cases have been traditionally treated with an open approach; nonetheless, as more experience and technology are available, a laparoscopic or robotic technique can be feasible in most patients [13,15]. In our patient, a laparoscopic approach aided to achieve a safer vision of the vascular structures around the mass.

Radiotherapy can also be used in selected cases where invasive surgery can be complex or dangerous. However, it's been associated with many complications and high recurrence rates. Neoadjuvant radiotherapy has been proved to be feasible in previous unresectable UCD [12,14]. In any case, surgery is recommended and necessary for a differential diagnosis in UCD [10]. Complete resection is advantageous as recurrences are rare, yet careful observation and follow-up are needed.

Conclusions

Abdominal UCD is a rare and still poorly understood pathology, but it must be considered when approaching a patient with an abdominal mass. As variable presentations are common, high clinical awareness and prompt diagnosis are of paramount importance. Surgery appears the most effective treatment, and with the advent of laparoscopy, better results with fewer complications are possible as we can perform a precise and bloodless dissection in areas of major vascular structures.

Patient perspective

At first, the patient was unsure about his treatment, how long it would last, whether it would hurt, and whether she could be "cured"; nonetheless, since surgery was successful, he was grateful to the medical team and was thankful that the cancer was gone.

Ethical approval

This article does not contain any studies with human participants or animals performed by any of the authors, and has been approved by the ethics committee or our hospital.

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None.

Author contribution

FS analyzed and interpreted the patient data. MC and FS were a major contributor in writing the manuscript. GM, AY and EF revised the manuscript and reviewed all the available data, All authors read and approved the final manuscript.

Consent

We declare no conflict of interest.

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Guarantor

Gabriel Molina MD, Iess Q. S. General Surgery, Quito Ecuador & Universidad San Francisco de Quito, USFQ, Quito Ecuador.

Declarations

- The submission has not been previously published, nor is it with another journal for consideration
- All authors listed on the paper have reviewed and approved the manuscript in its current form and are aware that it has been submitted.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Declaration of competing interest

We have no conflict of interest to declare

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2021.102494.

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