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Retroperitoneal Tumor: Castleman's Disease as a Differential Diagnosis - Case Report

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1. Abstract

1.1. Introduction

Castleman's disease (CD), described in 1956, is characterized as a low-incidence lymphoproliferative disorder with varied clinical manifestations, from asymptomatic to severe disseminated forms. The diagnosis is confirmed by histopathology and immunohistochemistry and should be considered after more common lymphoid diseases have been ruled out. This paper aims to describe a case of CD as a retroperitoneal tumor.

1.2. Case Report: A 45-year-old asymptomatic female patient was diagnosed with a retroperitoneal tumor by computed tomography. The image showed a solid, hypervascular nodular formation in the hepatopancreatic space, suggestive of a neoplastic lesion, with paraganglioma being the main diagnostic hypothesis. Surgical exploration was performed and was found to have a cystic tumor measuring around 5 cm, located in the retroperitoneum, in close contact with the proper hepatic artery and the head of the pancreas. Histopathological analysis revealed a lymph node structure with exuberant parafollicular hyperplasia and an onion-skin appearance. These findings were indicative of Castleman's disease, confirmed by immunohistochemistry.

1.3. Conclusion: Castleman's disease is rare and should be considered as a differential diagnosis for retroperitoneal tumors. In this case report, the unicentric form was diagnosed and the lesion was completely resected.

2. Introduction

Castleman's disease (CD) is a rare and heterogeneous lymphoproliferative disorder described by Benjamin Castleman in 1956 [1], also known as angiofollicular or giant lymph node hyperplasia. The most prevalent site of involvement is the mediastinum, followed by the cervical and abdominal regions [2]. The etiopathogenesis is mainly related to inflammatory and autoimmune states, as well as viral infections such as human immunodeficiency virus (HIV) and human herpesvirus type 8 (HHV-8) [3]. It mainly affects patients aged between 30 and 50, regardless of gender. The clinical presentation is non-specific and there are no biomarkers or definitive radiological findings for the diagnosis, which is established by a histopathology and immunohistochemistry study [4]. CD is classified according to its distribution in the human body, manifesting as a localized (unicentric) or disseminated form, involving more than one anatomical site (multicentric). It is also divided by histological pattern into four main types: hyaline vascular, plasma cell, transitional type and stromal type [4,5]. The unicentric form typically manifests as isolated lymph node enlargement with no evident systemic symptoms, often diagnosed as an incidentaloma on imaging. On the other hand, the multicentric form is characterized by generalized lymph node growth, constitutional symptoms, anemia, thrombocytopenia, elevated PCR and visceromegaly, usually with a clinical course that is more aggressive [4,5]. The presentation as a localized retroperitoneal mass has several differential diagnoses with entities such as sarcoma, lipoma, lymphoma

and paraganglioma, all of which have variable prognosis.^{4,7} For its rarity, this report aims to describe a case of retroperitoneal Castleman's disease in a patient treated at the Oswaldo Cruz University Hospital (HUOC).

3. Case Report

A 45-year-old asymptomatic female patient with hypertension and type 2 diabetes mellitus was admitted to the HUOC in June 2022 to undergo resection of a retroperitoneal tumor diagnosed by computed tomography (CT).

The patient's past medical history included a diagnosis of right nephrolithiasis in September 2021, when she presented with recurrent low back pain and dysuria. After appropriate management, she remained asymptomatic and had no changes in the laboratorial tests. In addition to nephrolithiasis, abdominal ultrasound showed a pancreatic cystic image, anechoic and circular in appearance, with vascularization in its middle, measuring 3.8 x 3.0 cm and similar to vascular ectasia. Angiotomography of the abdominal aorta showed a hypervascular heterogeneous nodular formation measuring 3.2 x 2.6 cm, with well-defined limits and located in the hepatopancreatic area. It was in close contact with the hepatic artery, but with no apparent communication. This finding was suggestive of a neoplastic lesion, being paraganglioma the main hypothesis. Laparoscopic surgery was first performed, identifying a cystic tumor that measured 5 cm. It was located in the retroperitoneum, inferior to the caudate lobe of the liver and in intimate contact with the proper hepatic artery along with the head of the pancreas. It was decided to convert the procedure to laparotomy due to hard-to-control bleeding after dissection was attempted. Therefore the tumor pedicle was isolated and resected with no evidence of vascular damage. The patient's recovery was satisfactory and she was discharged from hospital three days after the surgical procedure.

Histopathological analysis revealed a lymph node structure with exuberant vascular and parafollicular hyperplasia, along with an onion-skin appearance, mural hyalinization and atrophy of germinal centers. These morphological findings were indicative of Castleman's disease. The diagnosis was confirmed by immunohistochemical analysis, which showed normal distribution of T and B lymphocytes, as well as absence of BCL 2 and HHV-8 (Table

1). The sample had no signs of malignancy and the findings were compatible with hyaline-vascular type of Castleman's disease.

4. Discussion

CD is an uncommon disease of the lymph nodes and related tissues. The pathogenesis is complex and still uncertain.⁶ Preoperative diagnosis is difficult due to the wide range of differential diagnoses and variable presentation, with unicentric (UCD) or multicentric (MCD) behaviour [6,7]. It is often incidentally detected by imaging tests carried out during the investigation of other conditions⁸, such as the case described in this report.

The incidence of UCD in the abdomen is very low, especially in retroperitoneal peripancreatic location [6]. It is common for there to be no abnormal clinical phenomena or laboratory findings. In contrast, MCD presents with lymph node enlargement, fever, fatigue, weight loss, anemia, thrombocytopenia and elevated PCR, with a more aggressive nature [9].

The findings on imaging tests are non-specific and radiological appearance is similar to a wide range of diseases, including neoplasms (lymphoma, sarcoma, paraganglioma, lipoma), reactive lymph node hyperplasia and inflammatory diseases such as tuberculosis and sarcoidosis [8]. Ultrasound usually reveals a homogeneous mass with low density, while angiotomography shows a solid hypervascular tumor [9,10]. The diagnosis is made by anatomopathological analysis. However, certain lymphomas have similar characteristics to CD and immunohistochemistry is often necessary for confirmation [8-11]. The most common histological type is hyaline-vascular variant, which presents follicular centers surrounded by hyaline tissue and interfollicular vascular proliferation. This conformation is presented in the case as an "onion skin" appearance (figure 3), when the mantle zone is surrounded by concentric layers of lymphocytes [8,9]. CD usually has a benign histologic nature and most patients presenting the unicentric form are considered cured after complete excision of the lesion, with a low recurrence rate [8-11]. Therefore, surgical resection for both diagnosis [9] and treatment of UCD is the recommended option [8]. The rarity of retroperitoneal cases determines the need for strict postoperative follow-up [9]. The prognosis will depend on the extent of affected tissues [8].

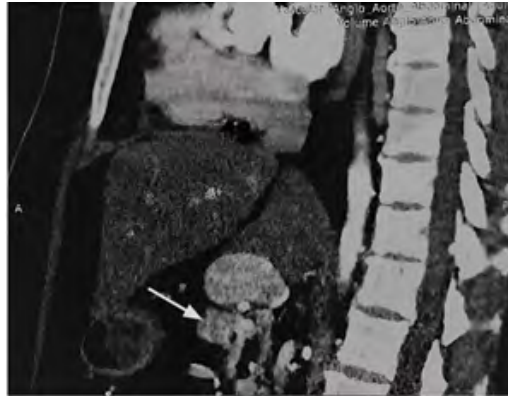


Figure 1: Angiotomography of the abdominal aorta showing a tumor (see arrow) with soft tissue attenuation, located inferiorly to the caudate lobe and superiorly to the pancreas.

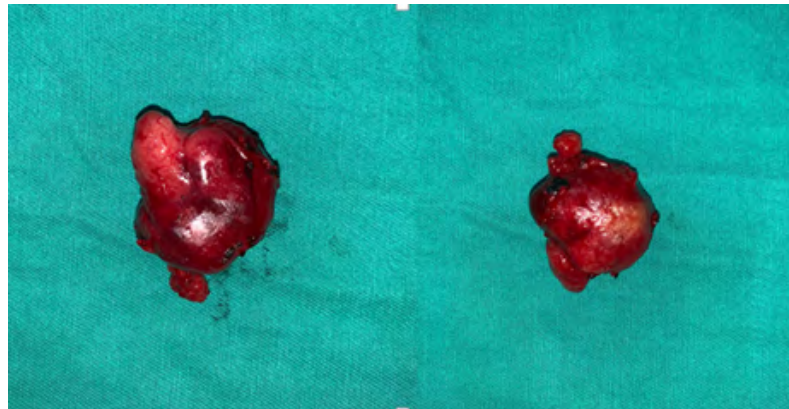


Figure 2: Retroperitoneal tumor after resection.

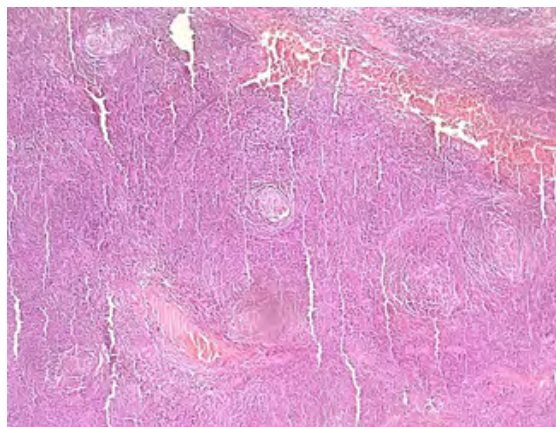


Figure 3: Lymphoid tissue showing an onion-skin appearance (optical microscopy, hematoxylin and eosin staining), characteristic of Castleman's disease.

Table 1: Immunohistochemical results showing negativity for BCL-2 and HHV-8

| Antibodies | Clone and Result |
|---|---|
| Ki- 67 - Cell proliferation antigen | M1B1 - Positive in central follicular cells |
| CD20 - B lymphocyte antigen | L26 - Positive in B cells |
| CD3 - T lymphocyte receptor (epsilon chain) | RBT-CD3 - Positive in T cells |
| CD10 - common antigen of acute lymphocytic leukemia (CALLA) | 56C6 - Positive in central follicular cells |
| Antiapoptotic protein BCL-2 | 124 - Negative in central follicular cells |
| Ciclina-D1 - Cell cycle regulating protein (BCL-1) | SP4 - Negative |
| HHV8 | LN-53 - Negative |

5. Conclusion

Castleman's disease should be considered as a differential diagnosis for retroperitoneal tumors. Immunohistochemical analysis of the surgical specimen defines the diagnosis. In this case report, the unicentric form was identified and the tumor was successfully resected.

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