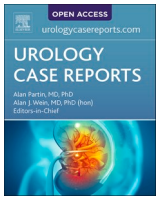
Urology Case Reports 57 (2024) 102849

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr



Oncology

Unilateral adrenal Castleman's disease: A case report

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ARTICLE INFO

Keywords:
Adrenal mass
Castleman's disease
Surgical resection

ABSTRACT

Castleman disease (CD) is a heterogeneous hematological condition characterized by distinctive histopathological features. The etiology remains unclear, and clinical symptoms are generally nonspecific. CD can occur in any location containing lymphatic tissue, with the mediastinum being the most common site, while adrenal involvement is rare. Adrenal CD is typically incidentally discovered during physical examination, commonly affecting one side. Imaging studies often make it challenging to differentiate from common adrenal tumors, necessitating pathological confirmation. Surgical intervention is the preferred treatment, and the prognosis is generally favorable.

This paper presents a rare case of a left adrenal mass, which was diagnosed as Castleman disease following surgical resection.

1. Introduction

Castleman disease (CD) is a heterogeneous hematological condition characterized by distinctive histopathological features, first described by Castleman in 1956, representing a complex lymphoproliferative disorder.[1] The etiology remains unclear, and clinical symptoms are generally nonspecific. CD can occur in any location containing lymphatic tissue, with the mediastinum being the most common site, while adrenal involvement is rare. Adrenal CD is typically incidentally discovered during physical examination, commonly affecting one side.[2] Imaging studies often make it challenging to differentiate from common adrenal tumors, necessitating pathological confirmation. Surgical intervention is the preferred treatment, and the prognosis is generally favorable.

This paper presents a rare case of a left adrenal mass, which was diagnosed as Castleman disease following surgical resection.

2. Case report

The patient is a 50-year-old male with no history of hypertension, diabetes, or previous abdominal surgeries. In March 2023, he was referred to our hospital after a left adrenal mass was discovered during a routine examination at a lower-level hospital. Upon admission, the patient underwent a complete abdominal CT scan, which revealed a left retroperitoneal mass measuring approximately 55mm × 40mm. The mass had well-defined margins, with multiple calcifications and mild enhancement observed (Fig. 1 A, B, C). Other imaging studies showed no lymphadenopathy or additional tumors. The patient underwent dynamic blood pressure monitoring, which indicated no hypertension. Additional laboratory tests, including complete blood count, biochemistry, coagulation, and myocardial injury markers, showed no significant abnormalities. Adrenal hormone assessments, including renin, aldosterone, 24-h urinary vanillylmandelic acid, catecholamines, ARR, and cortisol, were all within normal ranges.

After completing the evaluation, the patient underwent laparoscopic resection of the left retroperitoneal mass. Intraoperatively, the retroperitoneal tumor exhibited mild adhesion to surrounding tissues. The surgery was successful, and the resected specimen was light brown, firm, and measured 60mm × 42mm x 35mm. Upon sectioning, the tumor revealed a pale yellow tissue (Fig. 1 D). Postoperative pathological examination and immunohistochemical results showed: CD3 (++), CD20 (++), CD31 (+), CD34 (vascular +), KI-67 (10 % +), Vimentin (−), CD5 (+), Cyclin D1 (focal +), BCL-2 (non-germinal center +), and BCL-6 (germinal center +). Hematoxylin and eosin staining revealed small vessel proliferation and thickened vessel walls, with a central region containing transparent and sclerotic vessels surrounded by concentric layers of small lymphocytes (Fig. 1E and F). Based on the HE staining and immunohistochemical results, the final diagnosis was para-adrenal

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https://doi.org/10.1016/j.eucr.2024.102849
Received 6 August 2024; Accepted 11 September 2024
Available online 12 September 2024

Castleman disease (giant lymph node hyperplasia) on the left side. The patient was discharged smoothly 5 days post-surgery without complications. Three months after discharge, a follow-up abdominal CT revealed no tumor recurrence at the surgical site or other locations, and all laboratory tests showed no significant abnormalities, indicating a favorable surgical outcome.

3. Discussion

Castleman disease (lymphoproliferative disorder) is a rare benign lymphoproliferative condition. Epidemiological studies indicate an incidence rate of approximately 2125 cases per million, with a median age of onset between 34 and 40 years. Castleman disease (CD) can affect multiple locations, typically involving lymph nodes in the neck, abdomen, and mediastinum, with rare occurrences in the retroperitoneal adrenal region.[3] The pathogenesis of adrenal Castleman disease remains unclear; however, several theories exist, including abnormal expression of interleukin (IL)-6 and elevated levels of vascular endothelial growth factor (VEGF) associated with viral stimulation and angiogenesis. Due to the lack of non-specific clinical manifestations and imaging features, early diagnosis of adrenal Castleman disease is often challenging. Such cases are typically discovered during physical examinations, with definitive diagnosis reliant on pathological and immunohistochemical results.

Generally, large retroperitoneal and adrenal masses require assessment of adrenal-related hormones to aid in determining the nature of the tumor. In cases of adrenal Castleman disease, most do not present with significant adrenal insufficiency or excess hormone production.[4] The histological types of Castleman disease (CD) typically include hyaline vascular, plasma cell, and mixed variants. The hyaline vascular type is the most common, accounting for approximately 90 % of CD cases, characterized by abnormal lymph node follicles and vascular proliferation in the interfollicular areas, usually without specific clinical symptoms. Clinically, Castleman disease (CD) is classified into unicentric CD (UCD) and multicentric CD (MCD). UCD is the most common form, while MCD has a poorer prognosis.[5] On imaging, hyaline vascular type adrenal Castleman disease typically presents as a homogenous, well-defined soft tissue mass on CT. In contrast, plasma cell type UCD is less common, generally larger, and more likely to exhibit necrosis, peripheral enhancement, and retroperitoneal lymphadenopathy, indicating malignant features.[6] In this case report, the adrenal tumor presented on CT as a homogeneous, well-defined mass with mild enhancement and calcification, with no tumors noted in other areas. Related adrenal hormone levels showed no significant abnormalities. Postoperative pathological and immunohistochemical results suggest unicentric hyaline vascular type Castleman disease.

The treatment plan for Castleman disease (CD) should be determined based on the site of occurrence, tumor quantity, and pathological subtype, including options such as surgical intervention, chemotherapy, and targeted therapy. Surgical resection is the primary treatment option for adrenal Castleman disease (CD).[7] If the lesion can be completely excised, prognosis is generally favorable regardless of pathological subtype, clinical presentation, or immunohistochemical results, and no adjuvant therapy is typically required.[8] However, since a small subset of adrenal Castleman disease (CD) may present with catecholamine hormone abnormalities and hypertension, it is essential to preoperatively control blood pressure and heart rate using alpha-blockers in patients with these conditions.[9] Additionally, studies have reported an increased risk of obstructive bronchiolitis, AA amyloidosis, and lymphoma in patients with Castleman disease (CD). Therefore, adrenal CD patients with persistent abnormal clinical symptoms, even after complete tumor resection, should undergo regular follow-up.[10] Multicentric Castleman disease (MCD) is a more aggressive form of CD, typically presenting at an older age compared to unicentric Castleman disease (UCD). MCD is reported to be a potentially malignant condition with a generally poor prognosis. Although the optimal treatment approach remains undetermined, some success has been achieved with chemotherapy, systemic corticosteroids, radiotherapy, and combination therapies. However, the mortality rate remains high, approximately 50 %, indicating significant room for advancement in the management of MCD.[11]

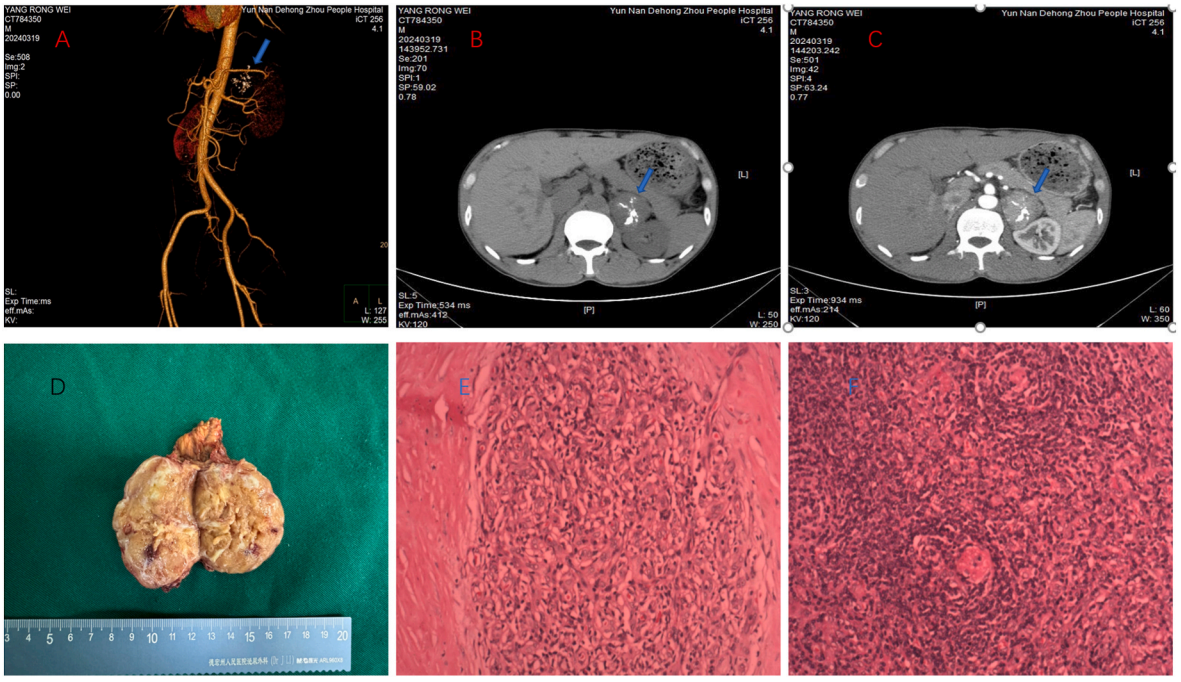

Fig. 1. Enhanced CT scans of the entire abdomen show a homogeneous, mildly enhancing mass in the left adrenal region, measuring approximately 55mm × 40mm, with accompanying calcifications and no surrounding enhancement (A, B, C). The resected specimen has an intact capsule, a light brown exterior, firm texture, measuring 60mm × 42mm x 35mm, and reveals pale yellow tissue upon incision (D). Hematoxylin-eosin (HE) staining shows proliferation of small blood vessels and thickening of vessel walls, with the center containing transparent and sclerotic vessels surrounded by concentric layers of small lymphocytes (E, F). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

4. Conclusion

Adrenal Castleman disease is a lymphoproliferative disorder occurring in the adrenal region, often difficult to differentiate from common adrenal tumors and lymphomas on imaging. Diagnosis is based on pathological and immunohistochemical findings. Surgical resection is the primary treatment modality with a favorable prognosis; however, regular follow-up is required for non-vascular type CD.

CRediT authorship contribution statement

Ji Li: Conceptualization. **Jing Bai:** Methodology. **Haifeng Wang:** Writing – review & editing. **Haole xu:** Writing – original draft.

Declaration of competing interest

The authors declare no conflict of interest.

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