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A case of adrenal Castleman disease combined with liver cirrhosis and splenomegaly

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ABSTRACT

To describe a rare case of left adrenal Castleman disease (CD), splenomegaly, and cirrhosis. An examination revealed a left adrenal mass for more than three months, the patient, 44, was well-prepared for surgery after her left adrenal tumor was removed laparoscopically using a retroperitoneal approach, her postoperative pathology suggested that she had Castleman disease of the adrenal glands, and there had been no metastasis or recurrence during the six-month follow-up period. We have evaluated linked literature reports in this article, reporting relevant clinical knowledge regarding the disease and synthesizing previous research, in an effort to increase our understanding of it.

1. Introduction

Castleman disease (CD), also referred to as large lymph node hyperplasia or angiofollicular lymphoid hyperplasia, is a rare disease that lacks typical clinical symptoms and has an unknown etiology1.[1] Castleman, a distinguished pathologist, was the first to report and name the disease.[2] In clinical terms, unicentric CD (UCD) is the term used to describe a single lymph node involvement, while multicentric CD (MCD) is the term used to describe multiple lymph node involvement. Multicentric CD can be classified into hyaline-vascular, plasma-cellular, and mixed categories based on the pathological findings.[3]The plasma-cellular type is the most prevalent pathological type in MCD, while the hyaline-vascular type is the most prevalent in UCD.[4] Plasma cell type is the defining characteristic of MCD.[4] The literature contains a limited number of national and international reports, with the majority of them being presented as cases. The disease can manifest in any region of the body, with the mediastinum being the most common location in approximately 70 % of cases. It can also manifest in other regions, including the neck, axillae, lungs, mesentery, retroperitoneum, and pelvis.[3] However, CD in the adrenal glands is exceedingly uncommon.[5] We report a rare case of adrenal Castleman disease combined with cirrhosis and splenomegaly and conduct a review of the pertinent literature for future clinical work, in light of our dearth of in-depth study of this disease.

2. Case presentation

The patient, a 44-year-old woman, was admitted to the Yantai Affiliated Hospital of Binzhou Medical College because of a left adrenal mass found on physical examination for more than 3 months. She suffered from cirrhosis for a year and chronic hepatitis B for two years. History of hypertension for more than 1 year, maximum blood pressure 180/100 mmHg, oral nifedipine extended-release tablets, blood pressure controlled to 150/90 mmHg, not regularly monitored. A 2.4 cm × 3.5 cm oval soft tissue lesion with significantly less homogenous density, poorly delineated adrenal gland, and noticeably inhomogeneous enhancement around the retroperitoneum is shown by abdominal scanning and enhanced CT (Fig. 1 A, B, C,D). Further complete blood routine, hypertension four, liver and kidney function, urinary 17 ketones, urinary catecholamines, methoxy epinephrine, methoxy norepinephrine, blood cortisol and other tests. Among them, white blood cells were 2.6*109/L, platelets were 80*109/L, blood potassium was 3.17 mmol/L, and the rest did not show any obvious abnormality. Pheochromocytoma was considered as a possibility for the left adrenal space-occupying lesion. Blood pressure was controlled with phenobenzamine hydrochloride tablets and stabilized at 120–130/80-90 mmHg, heart rate 80–90 beats/min. After completing the preoperative examination, transfusing 2 units of platelets and excluding relevant contraindications, laparoscopic left adrenal tumor resection was performed via

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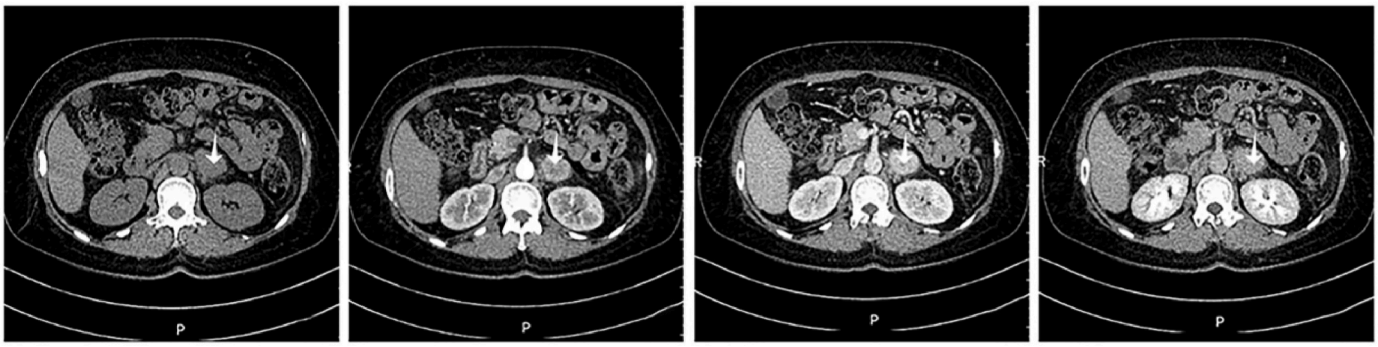

Fig. 1. CT plain view presentation 1A, CT arterial phase presentation 1B, CT venous phase presentation 1C, and CT delayed phase presentation 1D are the four types of CT presentations.

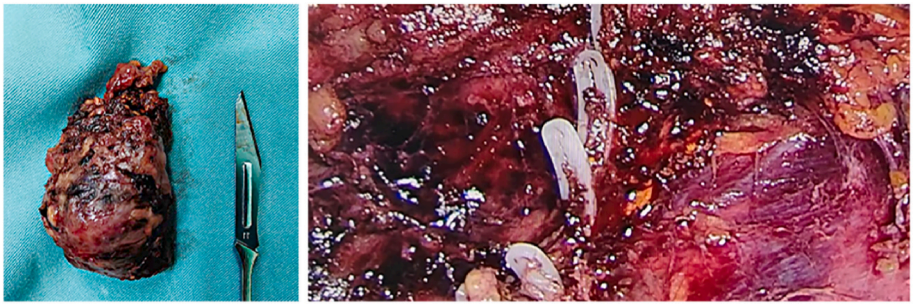

Fig. 2. A surgical specimen is in 2A, and a map of the operating area is in 2B.

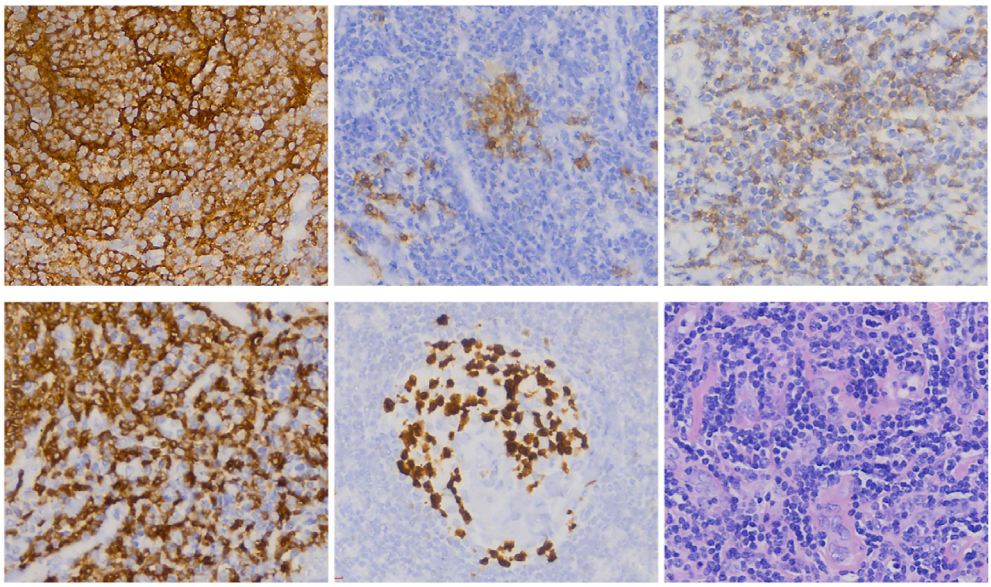

Fig. 3. CD21(+) represents 3A, CD38(+) represents 3B, and IgD (set area +) represents 3C, 4D is CD79a (+), 5E is Ki-67 (90 %+), and 6F is HE staining; (immunohistochemistry × 400x).

retroperitoneal approach, and the operation went smoothly(Fig. 2A and B). Postoperative pathologic diagnosis: Castleman's disease, hyaline vascular type. Immunohistochemistry (Fig. 3 A B C D E F): follicular zone CD20 (+), CD79a (+), germinal centers CD10 (+), BCL-6 (+), Ki-67 (about 90 % +), lgD (set of zones +), interfollicular zone CD3 (+), CD5 (+), BCL-2 (+), cyclinD1 (−), CD4 (+), CD8 (+), plasma cells CD38 (+), CD138 (+), FDC network CD21 (+), CD23 (+). After inviting the hematology department for consultation, no abnormality was found in the improvement of immunoglobulin, interleukin-6, β2 microglobulin, immunofixation electrophoresis, flow correlation CD series test, bone marrow smear cytology, neutrophil alkaline phosphatase staining, and WHO bone marrow chromosome karyotype analysis. The comprehensive diagnosis was unicentric CD (UCD), and the patient recovered well after surgery and was discharged 9 days after surgery. The patient did not see recurrence of metastasis in outpatient follow-up abdominal CT and laboratory tests six months after surgery, and was in good general condition.

3. Discussion

CD is an uncommon lymphoproliferative disorder that can manifest in individuals of all ages. Castleman's disease patients were identified in a study, with a mean age of 52.4 years. Of these patients, 56.1 % were male and 43.9 % were female.[1]Typically, CD is linked to infections with the cytokines interleukin-6 (IL-6), human herpesvirus 8 (HHV-8), and human immunodeficiency virus (HIV).[6] The etiology and pathogenesis of CD are not well understood. The majority of patients with UCD do not experience any ancillary symptoms, and a small number of them experience lymph node compression.[7] In addition to multiple enlarged lymph nodes throughout the body, patients with MCD frequently manifest with fever, anemia, abdominal distension and diarrhea, and hepatic and renal damage.[8] Additionally, the clinical manifestations of MCD are correlated with the serum HHV-8 viral burden, indicating that there may be a direct correlation with viral replication.[1]TAFRO syndrome is a unique subtype of MCD that may manifest as anemia, edema, fluid retention, renal insufficiency, hepatosplenomegaly, and thrombocytopenia.[1]After enhancement, the foci are significantly enhanced in the arterial phase and slightly attenuated in the venous phase, and CD is isointense in the CT scan. The lesion's enhancement was marginally diminished during the venous phase, but it continued to intensify. On magnetic resonance imaging (MR), it exhibits isosignal in the T1 sequence, a higher signal in the T2 sequence, and a clear progressive enhancement following enhancement scanning.[9] Research has demonstrated that the histologic type of Castleman's disease is closely associated with the CT imaging manifestations of the disease.[10] While imaging is instrumental in the differentiation of adrenal masses, it is not the sole determining factor.[8]The final diagnosis of CD is based on immunohistochemistry and examination. The hyaline-vascular type is primarily observed microscopically as hyperplastic lymphocytes arranged in concentric circles around atrophic germinal centers, while the plasma cell type is observed as clusters of plasma cells with varying degrees of plasma cell growth. The mixed type combines some of the structures of both types.[11] The disease is challenging to diagnose preoperatively and is susceptible to misdiagnosis. Consequently, it is necessary to distinguish it from pheochromocytoma, paraganglioma, multiple myeloma, and lymphoma. The preferable treatment option for the majority of patients with UCD is complete surgical resection of the lesion, which has been shown to effectively reduce the recurrence rate.[12] Additionally, a 3-year disease-free survival rate of 95 % has been reported.[3] The treatment of MCD is comparatively complex and has a poorer prognosis due to the diverse manifestations of MCD patients.[13] Consequently, the standard treatment options to date are still being explored. Glucocorticoids or other immunomodulators are combined with IL-6 monoclonal antibody or CD20 monoclonal antibody in the recommended treatment regimens at home and abroad.[8,14] Secondary or combined with malignant lymphoma necessitates appropriate combination chemotherapy.[15] The following reflections are made upon reviewing the diagnosis and treatment process: The patient's laboratory examination results are not typical, and the mindset of habitual thinking considered the possibility of pheochromocytoma, as well as the lack of understanding of Castleman's rare disease. The patient is in the loss of cirrhosis, with a history of splenomegaly, hypersplenism, and thrombocytopenia, and multicentric Castleman disease has similar manifestations. To avoid delay, the clinical type of Castleman disease should be clarified. The patient was diagnosed with decompensated liver cirrhosis, which was accompanied by splenomegaly, hypersplenism, thrombocytopenia, and other medical history. Multicentric Castleman disease exhibits comparable symptoms. Therefore, a more comprehensive examination is necessary to determine the clinical type of Castleman disease and prevent the condition from being postponed.

4. Conclusion

In conclusion, Castleman's disease is a rare disease that necessitates multidisciplinary collaboration for diagnosis. The possibility of this disease should be taken into account when encountering nonfunctioning adrenal masses with lymphadenopathy, as well as in patients with splenomegaly and thrombocytopenia. In addition to the consideration of common diseases such as cirrhosis,we should be vigilant for the possibility of this disease in order to prevent missed or misdiagnosis.

CRediT authorship contribution statement

Qilang Deng: Writing – original draft. **Yinglei Wang:** Resources. **Tong Liu:** Software. **Zhigang Li:** Investigation. **Bo-You Liu:** Methodology.

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