

Case Report

Retroperitoneal laparoscopic management method of Castleman's disease in the adrenal gland: two cases report and review of the literature

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Abstract: Purpose: Castleman's disease (CD) is a rare and complex disease of lymphoid tissues typically involving a mediastinal mass. CD in the adrenal area is an even rarer occurrence. In the present study, two extremely rare cases of adrenal Castleman's disease at our hospital are reported, and the relevant literatures were reviewed. Significant findings: A 51-year-old woman had abdominal pain for 1 month. Physical examination revealed a mass in the left abdominal. A computed tomography (CT) scan confirmed the presence of the mass. Additionally, a left suprarenal mass was detected in a 56-year-old male patient during a regular medical checkup. He had no symptoms when he arrived at our hospital. The two patients underwent mass excision via a retroperitoneal laparoscopic approach. Post-operative histopathological examination of both patients' specimens suggested a diagnosis of the hyaline vascular-type of CD. Conclusions: These two rare cases confirm that CD can occur in the adrenal gland area. In addition, we also demonstrate that retroperitoneoscopic surgical management is effective in the treatment of the disease.

Keywords: Castleman's disease, retroperitoneal laparoscopic, adrenal gland

Introduction

Castleman's disease (CD) is a rare disorder characterized by benign lymphoid proliferative neoplasm that was first described by Castleman in 1954 [1]. These neoplasms can be found in any part of the lymphoid tissues but are most commonly located in the chest, particularly in the mediastinum [2]. A search for relevant literature in the PubMed database using the following search strategy reveals that very few reports of CD in the adrenal glands exist, suggesting that CD of the adrenal glands is a comparatively rare occurrence: (Castleman's disease or giant lymph node hyperplasia) AND (adrenal or retroperitoneal). Below, we report two cases of CD in the adrenal gland, which were successfully managed using a retroperitoneal laparoscopic approach at our institution.

Case report

Case 1

A 51-year-old female patient presented with a 1-month history of abdominal pain, with no rel-

evant past medical or family history. Ultrasonography performed at another hospital revealed an 11 cm mass in the left adrenal area. Based on the ultrasonography findings, the patient received treatment in our hospital on an outpatient basis. Routine medical examination revealed a left abdominal mass with deep palpation. Results of routine blood tests, biochemical tests, and urinalysis showed no apparent abnormalities. An enhanced computed tomography (CT) scan revealed a 90 × 80 mm mass in the left adrenal area (**Figure 1A**).

After preoperative preparation, the mass was successfully removed using a retroperitoneal laparoscopic surgical approach. Postoperative histological examination of the resected specimen suggested a diagnosis of hyaline vascular-type CD. CD20 immunohistochemical staining revealed a closely follicular dendritic meshwork of cells (**Figure 1B**) and an "onion-skin" appearance characterized by concentric layers of small lymphocytes (**Figure 1C**). The operation was carried out without complications, and the patient has been without recurrent masses for 4 years.

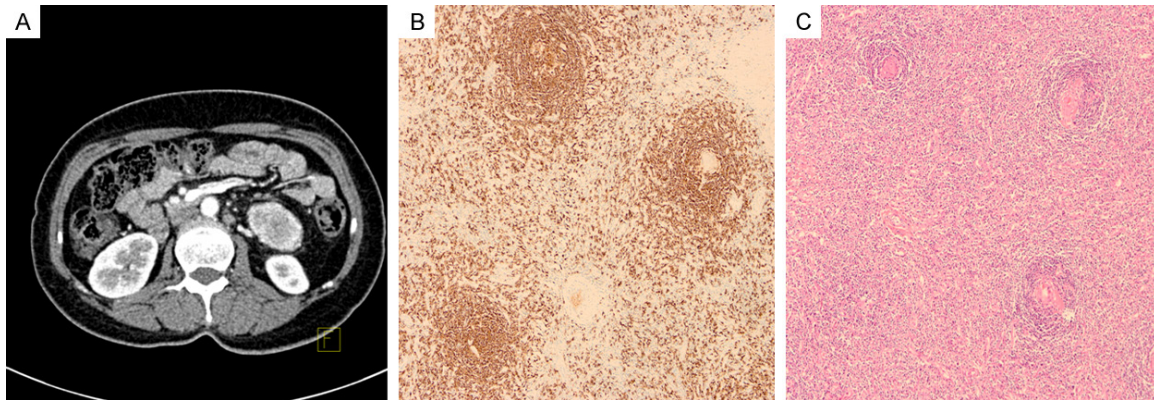


Figure 1. A. Preoperative computed tomography (CT) of patient 1: a mass in the left adrenal gland area. B. CD20 immunohistochemical staining (original magnification of $\times 100$). C. "Onion-skin" appearance characterized by concentric layers of small lymphocytes (Hematoxylin-eosin staining, original magnification of $\times 200$).

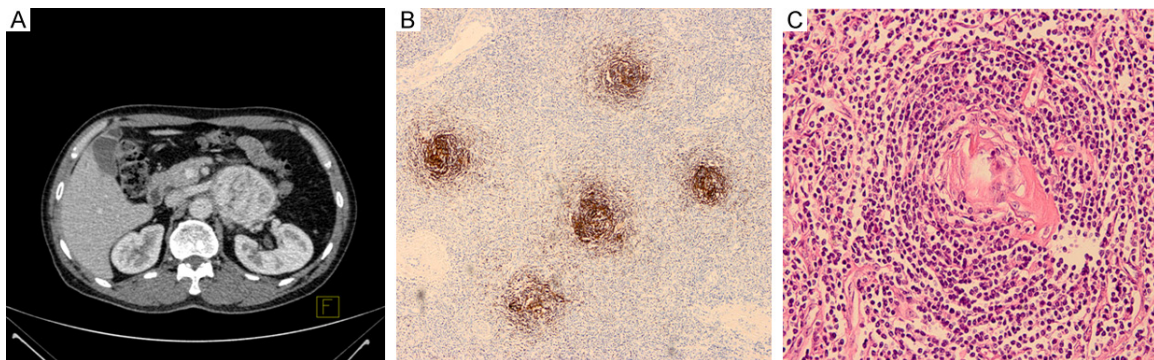


Figure 2. A. Preoperative computed tomography (CT) of patient 2: a mass in the left adrenal gland area. B. CD21 immunohistochemical staining (original magnification of $\times 100$). C. High-power microscopy showed that the germinal centers were composed of a hyalinized vessel and small lymphocytes arranged in a concentric circle pattern (Hematoxylin-eosin staining, original magnification of $\times 400$).

Case 2

A 56-year-old male patient was found to have a mass in his left suprarenal area by abdomen ultrasonography following a regular medical checkup 6 days prior. The patient had no discomfort. Physical examination revealed no relevant findings. Physiological indices including blood and urine routines, in addition to liver and renal function, did not reveal any noticeable abnormalities. He had neither a past medical history nor a family history suggesting any particular cause. An enhanced CT scan revealed a mass of approximately 80×58 mm in diameter with heterogeneous density (**Figure 2A**).

After sufficient preoperative evaluation, the tumor was managed by a retroperitoneal laparoscopic surgical method. Surprisingly, the histopathology also indicated hyaline vascular-type CD. The operation was performed without

complications and the patient walked out of the hospital one week after surgery. Immunohistochemical staining was positive for CD20 lymphocytes in the interfollicular zones (**Figure 2B**). High-power microscopy showed that the germinal centers were composed of a hyalinized vessel and small lymphocytes arranged in a concentric circle pattern (**Figure 2C**). After 3 years of follow-up, the patient was well and without any signs of disease.

Discussion

CD is an uncommon, chronic lymphoproliferative disorder that is characterized by benign proliferation of the lymphoid tissues [1]. It is otherwise known as giant lymph node hyperplasia, angiofollicular lymph node hyperplasia, angiomatous lymph node hamartoma, benign giant lymphoma, and follicular lymphoreticuloma [3-6]. CD mainly occurs in young people

between the ages of 15 and 35, while gender does not differentially predispose people to developing the disease [7]. It may occur in other lymph nodes of the body, although research shows that most CD occurs in the mediastinum. In addition, CD seems to occur in regions outside the lymphatic system in about 5% of cases [8]. It is quite unusual that both cases at our institution occurred in the adrenal area.

The exact etiology and pathogenesis of CD are still unclear. Some researchers believe that it related to human herpesvirus-8 (HHV-8) [9]. Aberrant production of interleukin (IL)-6, secondary to stimulation by a chronic infection of HHV-8 and subsequently leading to plasma cell proliferation, has been considered as a possible mechanism of CD [7, 10]. Additionally, human immunodeficiency virus (HIV)-associated CD is considered to be strongly correlated to HHV-8 [10]. In our two cases, both patients were HIV negative.

Clinically, CD is divided into two types: unicentric (UCD) and multicentric (MCD). The clinical symptoms of CD are not uniform, and vary between UCD and MCD [7, 8]. UCD usually appears in the lymphonodular regions either with the absence of symptoms, or with symptoms due to local compression, such as postprandial discomfort, vomiting and lumbar or abdominal pain [2, 7]. MCD is associated with symptoms and laboratory abnormalities due to the involvement of several lymph node stations and organs, such as anemia, thrombocytopenia, hypoalbuminemia, increased erythrocyte sedimentation rate, and so on [11]. In case 1, the patient had a history of abdominal pain, and these symptoms might due to pressure by the mass. However, the patient in case 2 had no clinical symptom. In fact, the clinical symptoms of CD are in general extremely various, and diagnose the disease through clinical presentations is difficult.

Histologically, CD is divided into three types: hyaline-vascular (HV), plasma cell variant (PC), and mixed-type [12]. As in our two cases, the HV type is characterized by giant lymph follicles centered on a central vessel, with an "onion skin" appearance. This is the most common type, and approximately 90% of all localized forms are of this type. However, the PC type is characterized by the infiltration of many more

polyclonal plasma cells. The PC type is a less common type of CD, and is associated with abnormal hematologic values and systemic symptoms. Mixed-type has feature of both of the above [13].

Generally, clinicians perform imaging investigations such as ultrasonography and CT at the location of physical symptoms. However, because of the lack of specific imaging features, CD is usually not considered as the first diagnosis [14]. Therefore, it is also hard to diagnose the disease by imaging tests before operating on the patient. Histopathological examination is necessary for the accurate diagnosis of CD.

A review of the literature concerning CD reveals that complete surgical resection may be the best treatment option for UCD. If the operation is performed without complications, the medical prognosis is very promising and no further treatment is needed. However, incomplete resection may lead to worse outcomes [15]. Unfortunately, the prognosis for MCD is significantly poorer. The role of surgery is limited to obtaining tissue for biopsy and to debulking the mass to address specific problems such as bowel obstruction, vascular or airway compromise, and massive organomegaly [15, 16]. Chemotherapy and immunotherapy may be needed to treat multisystem involvement. Dan et al. reported that UCD in the pararenal retroperitoneum could be treated by a laparoscopic approach [17]. In our two cases, we successfully removed the masses by retroperitoneal laparoscopic method. We think the approach also may offer magnified images to facilitate and secure dissection.

In conclusion, CD is a benign, lymphoproliferative disease that rarely occurs in the adrenal gland area. CD should be considered during differential diagnosis in the adrenal area. In addition, the retroperitoneal laparoscopic approach demonstrated in the present study may be an appropriate treatment method for CD in the adrenal gland area. This minimally invasive treatment has many advantages, such as very little trauma and rapid recovery.

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Disclosure of conflict of interest

None.

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