

# Abdominal giant lymph node hyperplasia (Castleman's disease): report of two cases

## Case Report

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**Abstract:** **Objective:** To describe the clinical characteristics and diagnostic features of giant lymph node hyperplasia or Castleman's disease found in rare locations. **Methods:** Two cases of Castleman's disease (1 abdominal and 1 retroperitoneal) were confirmed histopathologically. The clinical and medical imaging features were described and relative literatures were reviewed. **Results:** One patient had no clinical symptoms and the other had epigastric discomfort. The location of the benign tumor was retroperitoneal and left adrenal gland, respectively. Both cases were of solitary tumor, 1 was a hyaline-vascular type and the other was a mixed type with plasma cell and hyaline-vascular type. Both were successfully treated by surgical excision. Patients were followed up for 3 and 4 years respectively with no signs of recurrence on CT imaging. **Conclusions:** Abdominal Castleman's disease lacks specific clinical manifestations. Definitive diagnosis requires histologic examination and excisional surgery is the method of choice for treatment.

**Keywords:** Giant lymph node hyperplasia • Castleman's disease • Diagnosis • CT imaging • Surgery

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## Introduction

Castleman's disease, also known as angiofollicular lymph node hyperplasia, giant lymph node hyperplasia, and angiomatous lymphoid hamartoma, is a rare nonneoplastic lymphoproliferative disorder of unknown etiology. It was first reported by the Castleman in 1956 and was subsequently named Castleman's disease [1]. Histopathological examination of the enlarged lymph nodes often shows a follicular hyperplasia of lymph nodes with abnormally increased interfollicular vascularity. This disease usually occurs in the chest, especially in the mediastinum and neck [2-6], and rarely occurs in the abdomen. The usual presentation is a solitary and well-circumscribed asymptomatic mass lesion, often be-

coming large size, with infrequent hematologic manifestations [2-6]. We report 2 cases of unusual Castleman's disease located in the abdominal and retroperitoneal cavities. The clinical and radiological features of these rare types of Castleman's disease are described.

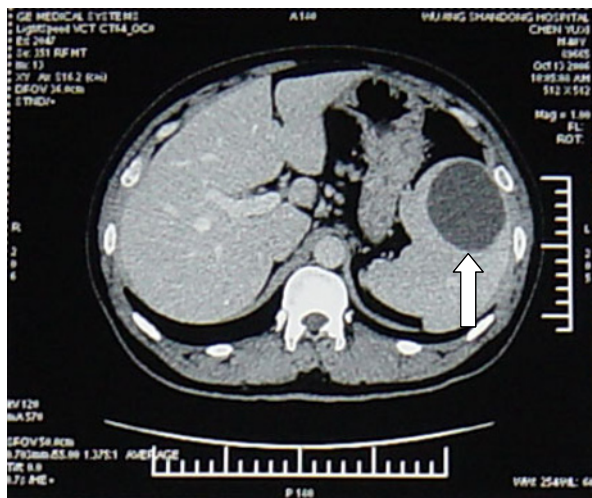
## Case reports

Case 1. A 39 years old male subject was admitted to our hospital in October 2006 for a splenic cyst and a retroperitoneal solid mass identified by ultrasound examination during a routine annual health exam. He had a 10-year history of Wolf-Parkinson-White syndrome. He exhibited no other symptoms except frequent com-

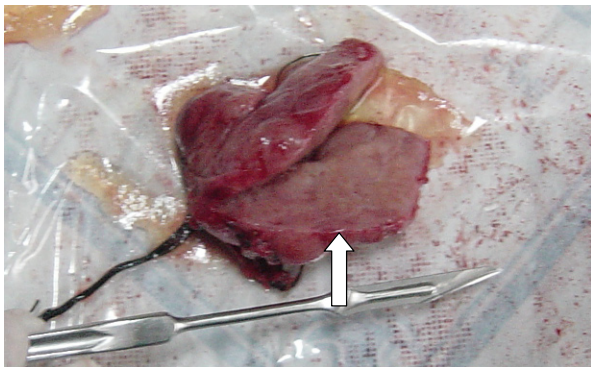
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plaints of “coldness.” Physical examination was unremarkable and there was no palpable mass in the abdomen. Blood tests and chest X-rays were normal. CT scan showed a 7cm × 8cm × 6cm mass attached to the spleen (Figure 1). Laparotomy was performed under general anesthesia. A 5cm × 6cm × 7cm cystic mass was seen on the upper pole of the spleen. The mass was dumbbell-shaped and capsulated (Figure 2). Histopathological examination confirmed Castleman’s disease of hyaline-vascular type. Clinical examination and CT abdominal imaging 4 years after the surgery showed no signs of recurrence.

Case 2. A 65 years old male presented with upper abdominal discomfort and pain for 3 days. Abdominal CT scan revealed a solid lesion on the left adrenal gland. He was admitted on 7 October, 2007. Physical examination of the abdomen was unremarkable and blood cancer markers (alpha fetoprotein, carcinoembryonic antigen, carbohydrate antigen 19-9) were negative. Under general anesthesia, laparotomy was performed, and a 3cm × 3cm × 2cm solid tumor was found on the



**Figure 1.** Case 1, CT showed a 7cm × 8cm × 6cm low-density mass on the upper pole of the spleen (arrow)



**Figure 2.** Resected specimen from case 1.

left adrenal gland. The tumor was completely resected. Histopathological examination of the tumor confirmed as Castleman’s disease, with a mixture of plasma cells and hyaline-vascular type. He was followed up for 3 years. Clinical examination and CT abdominal imaging showed no signs of recurrence.

## Discussion

There is no remarkable sex predominance or identifiable risk factor for the development of Castleman’s disease [3]. Based on clinical and radiological findings, Castleman’s disease can be classified as unicentric and multicentric [7]. Unicentric Castleman’s disease is usually a slow growing solitary mass typically located in the mediastinum or mesenteries, with no constitutional symptoms and no elevation of acute phase reactants, such as interleukins, erythrocyte sedimentation rate or C-reactive protein. Patients are usually either asymptomatic or have nonspecific symptoms, such as coughing, dyspnea, chest pain, respiratory infection, and back pain, mainly caused by tracheobronchial compression [8]. Multicentric Castleman’s disease affects more than a single group of lymph nodes, with widespread lymphadenopathy and in some instances hepatosplenomegaly [7]. This form sometimes occurs in those infected with human immunodeficiency virus (HIV). Multicentric Castleman’s disease runs a more aggressive course and can progress to non-Hodgkin’s lymphoma that requires systemic therapy. This type of Castleman’s disease is often associated with severe fatigue, night sweats, fever, weight-loss, anorexia, due to overproduction of interleukin-6 [7]. Castleman’s disease can also be classified into hyaline-vascular type and plasma cell type, based on a combination of various histological features [2,3]. The more common hyaline vascular subtype is characterized by small hyaline vascular follicles and interfollicular capillary proliferations. The present report described two unusual cases of unicentric Castleman’s disease where a single enlarged lymph node was found in the retroperitoneal cavity or on the left adrenal gland. Histopathological examination confirmed one case was of hyaline-vascular type and the other was a mixed type of hyaline-vascular and plasma cell type. One of the patients had non-specific upper abdominal aches before imaging examination. The second patient was asymptomatic and the benign tumor was discovered during a routine abdominal ultrasound exam. These results suggest that unicentric Castleman’s disease located in the abdominal or retroperitoneal cavities are often asymptomatic and clinical diagnosis can be difficult without imaging examinations.

Currently there are no standard diagnostic criteria for Castleman's disease and the initial diagnosis relies on a high index of suspicion. There has been no evidence to show that any particular group of population is more susceptible to this disease therefore mass screening of this rare condition is unhelpful. An enlarged lymph node, usually inside the chest or abdomen, is often the only abnormality for most people with the localized form of Castleman's disease. In patients with systematic symptoms or non-specific symptoms, such as cough, shortness of breath, trouble eating, abdominal pain, or just a feeling of fullness, diagnosis may be based upon a thorough clinical evaluation that includes a detailed patient history and a variety of specialized imaging techniques. The diagnosis can be made initially on medical imaging but confirmation of diagnosis requires histopathological examination of the surgical specimen. Plain chest X-ray may reveal mass lesions in the mediastinum, hilar lymph node enlargement, widened mediastinum, bilateral pulmonary nodules or pleural effusion [9]. Ultrasound examination may reveal lesions in the neck, abdomen, retroperitoneal and pelvis. CT and MRI scan are also useful in localizing the mass, as localized Castleman disease manifests as either a solitary, well-circumscribed mediastinal mass or an infiltrative mass with associated lymphadenopathy on CT or MR images [9]. PET-scanning can complement CT-scanning on the metabolic status of lymph nodes.

Treatment of unicentric Castleman's disease is largely surgical and post-operative prognosis is excellent. Up to 95% of the patients with unicentric Castleman's disease are cured with surgical resection and the prognosis is excellent with a 5 yr survival of close to 100% [5]. In a re-

cent review of 13 patients, 12 underwent excisional surgery [10]. After a mean follow-up of 63.3 months, only one patient showed recurrence, but this patient remains progression-free 7 years after repeat resection [10]. The two patients in the present report have been followed up for 3 and 4 years, respectively, following the surgery, and clinical examination and CT scans showed no signs of recurrence. Treatment for multicentric Castleman's disease has been challenging and there are no standard therapeutic regimens today. As the disease involves lymph nodes at multiple locations, surgery is mainly used for the diagnosis and for debulking the disease in some patients [11]. Corticosteroids, chemotherapy, radiotherapy and immune therapy have been helpful [11, 12]. Interferon (IFN)-alpha, rituximab, anti-IL-6 receptor antibodies, and thalidomide have been used in the management of multicentric Castleman's disease with some success [11,13,14]. Patients with multicentric Castleman's disease are at increased risk for developing frank malignant lymphoma such as non-Hodgkin lymphoma or Kaposi sarcoma.

In conclusion, Castleman's disease is a rare disease involving single or multiple lymph nodes. Unicentric Castleman's disease is often found in the chest, but on rare occasions it can be found in retroperitoneal cavity or adrenal glands. Diagnosis is based on clinical examination and medical imaging, but definitive diagnosis requires histopathological examination. Excisional surgery is the method of choice for treatment for unicentric Castleman's disease. Management of multicentric Castleman's disease is likely to require a combination of chemotherapy, radiotherapy and immune therapy.

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