

**Case Report**

## A Case of Unicentric type Castleman's Disease in the Retroperitoneal Space

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### Abstract

We herein present a case of unicentric type Castleman's disease (CD) arising in the retroperitoneal space. An asymptomatic 44-year-old male was noted to have an abdominal mass during his annual health checkup. The only abnormal finding on laboratory testing was an increased erythrocyte sedimentation rate. Preoperative imaging studies led to a possible diagnosis of unicentric type CD in the retroperitoneal space, and para-aortic lymph node swelling. Upon surgical exploration, the tumor, which measured 8cm × 5cm × 4cm, was obscured in the retroperitoneal space by the severe chronic inflammation of the surrounding tissue. The tumor, which had an abundant blood supply, was excised from the surrounding tissue, while the para-aortic lymph node was left intact. The histopathologic examination revealed that the tumor was comprised of hyaline vascular type tissue. His postoperative course was uneventful, and a postoperative CT scan performed 8 months after surgery showed no recurrent tumor, and marked shrinkage of the lymph nodes.

(Key words: Castleman's disease, unicentric type, retroperitoneal tumors)

### I. Introduction

Castleman's disease (CD) is a rare lymphoproliferative disorder that may develop in the lymph node tissue throughout the body. As in the first description by Benjamin Castleman, which characterized mediastinal masses, the tumor usually presents in the thorax and neck of young adults<sup>1</sup>. We present the case of patient with a solitary tumor, with concomitant lymph node swelling, arising in the retroperitoneal space.

### II. Case Report

A 44-year-old male without symptoms underwent his annual health checkup, and follow-up examination for a gallbladder polyp. Ultrasonographic study revealed a tumor in the abdominal cavity, and he was referred to our hospital for further examination and treatment.

Upon hospitalization, the physical examination on the afebrile patient revealed a mass in the left abdomen, near the umbilicus. Palpatory examination revealed a nontender immobile abdominal mass. Laboratory analysis revealed an increased erythrocyte sedimentation rate (ESR) of 17mm/hr, and a normal

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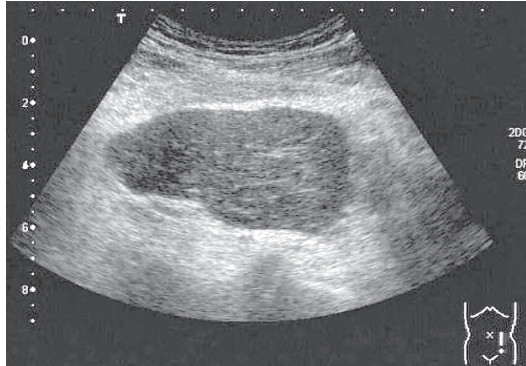


Fig. 1 Ultrasonographic study of the abdomen

The ultrasonographic study of the abdomen reveals a well-defined round mass, measuring 6.5cm in its maximal diameter, in the left retroperitoneal space. The parenchyma of the tumor is diffusely homogeneous with scattered tiny high echoic spots.

white blood cell count, C-reactive protein (CRP) level, and serum level of interleukin 6 (IL-6). Tumor markers, including the carcinoembryonic antigen (CEA), the carbohydrate antigen 19-9 (CA19-9), and the prostate-specific antigen (PSA) were not elevated. Antibodies to the human immunodeficiency virus and to the human T-cell lymphotropic virus type 1 were negative.

Ultrasonographic study of the abdomen revealed a well-defined round mass, measuring 6.5cm in maximal diameter, in the left retroperitoneal space adjacent to the left common iliac artery. The parenchyma of the tumor imaged as diffusely and homogeneously low echoic, with scattered tiny internal high-echoic spots (Fig. 1). A computed tomographic (CT) study confirmed that the tumor was located in the retroperitoneal space, between the left iliopsoas muscle and the left common iliac artery. The tumor was well circumscribed, and the homogeneous density of the tumor was lower than muscle and higher than fat (Fig. 2-1). Following the intravenous injection of contrast material, a marked enhancement of the tumor



Fig. 2-1 Plain CT scan

A CT scan confirmed the tumor to be located in the retroperitoneal space, between the left iliopsoas muscle and the left common iliac artery. The parenchyma of the tumor has a homogeneous density that is slightly lower than that of muscle, and much higher than fat.

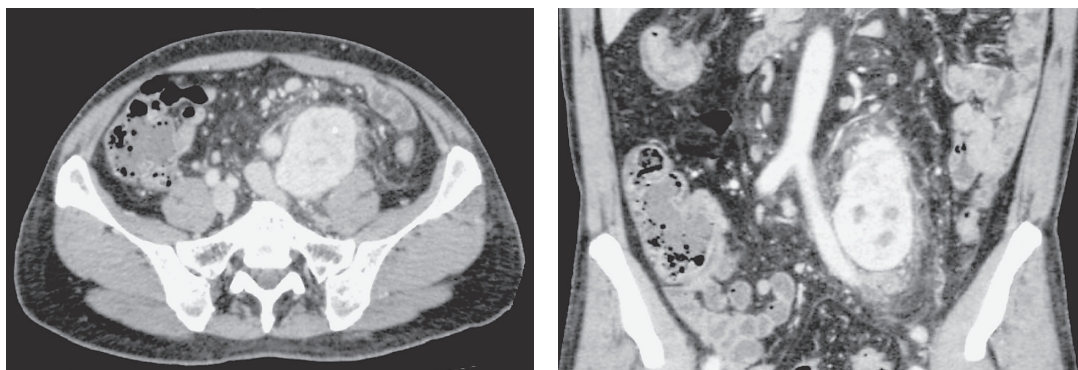


Fig. 2-2, 2-3 Enhanced CT scan

The tumor is well circumscribed, and the parenchyma of the tumor is strongly enhanced following contrast medium injection. There are some patchy areas that are not enhanced by the contrast medium.

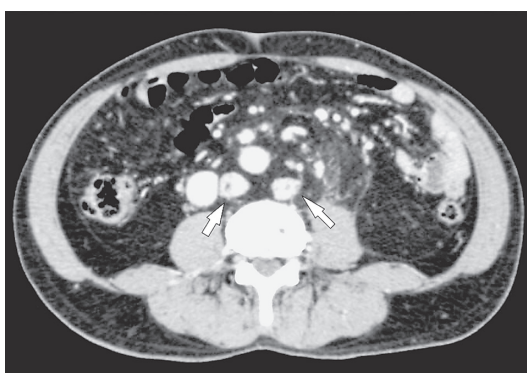


Fig. 3 Enhanced CT scan

There are some swollen lymph nodes in the retroperitoneal para-aortic space (arrows).

parenchyma was noted, with a non-enhancing, patchy internal area (Fig. 2-2, 2-3). Several swollen lymph nodes, of which the maximal diameter was 18 mm, were visualized in the retroperitoneal para-aortic space (Fig. 3). A gallium scintigraphic scan failed to reveal any abnormal uptake. The endoscopic examinations revealed no pathologic lesions in the stomach or the colon.

The differential diagnoses of the retroperitoneal tumor included liposarcoma, leiomyosarcoma, malignant fibrous histiocytoma (MFH), and malignant lymphoma. However, the internal density of the tumor was not identical to those of fat or muscle, and the parenchyma of the tumor showed no evidence of necrotic or cystic degeneration. Moreover, marked enhancement was noted after intravenous contrast injection. These findings, along with the results of a negative Gallium scan, prompted us to consider the diagnosis of CD. With reference to the absence of symptoms and the minimal hematologic abnormality, we considered that the tumor was a unicentric type CD, and that the swelling of the para-aortic lymph nodes was due to the response to the tumor presence.

At operation, the retroperitoneal tumor was found to be located behind the parietal viscera of the retroperitoneum, and dorsal to the ureter and the testicular artery. The retroperitoneal tissue around the tu-

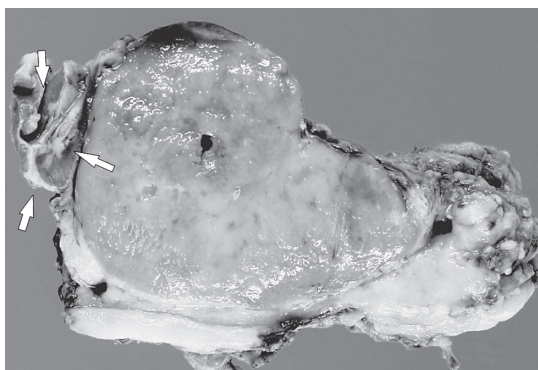


Fig. 4 Macroscopic findings

Macroscopically, the retroperitoneal tumor is well-circumscribed and measures 8 cm in diameter. The cut surface is homogenous and brown. The parenchyma of the tumor is elastic and hard, and is surrounded by a thin fibrous capsule. The early stage of the same histologic change is seen in the adjacent small lymph nodes (arrows).

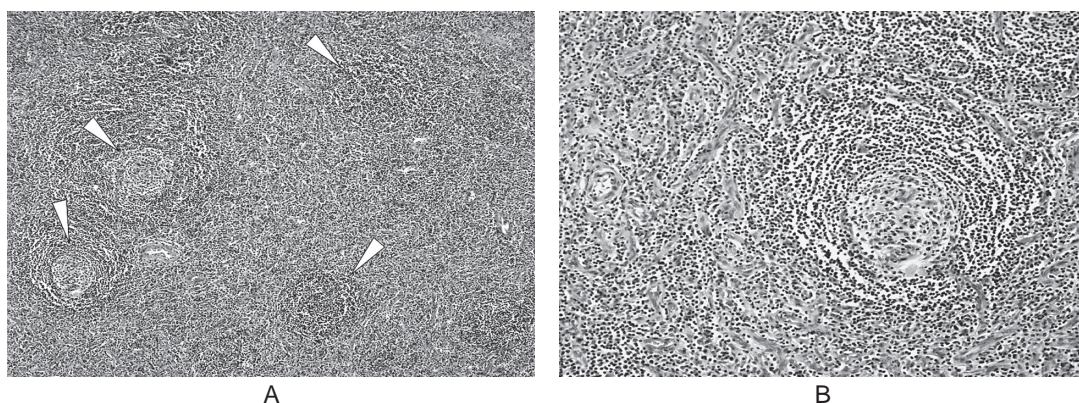


Fig. 5 Pathologic findings

Low (A) and high (B) magnified view of hematoxylin-eosin staining section. There are scattered lymphoid follicles (arrowheads), and sinuses are absent. Richly vascularized stroma is seen in and around the lymphoid follicles.

mor had become thickened and indurated due to chronic inflammation, and it obscured the tumor and the surrounding organs and tissues. The retroperitoneal tumor, which had an abundant blood supply from the surrounding tissue, was removed with the adherent iliopsoas muscle, the adjacent fat, and the surrounding tissue.

Macroscopically, the excised tumor was 8cm × 5cm × 4cm, and the cut surface was brown. The parenchyma of the tumor was elastic and hard, with no necrotic area and was surrounded by fibrous tissue (Fig. 4).

Histologically, scattered lymphoid follicles were found in the richly vascularized stroma, while sinuses were absent (Fig. 5). These characteristic histopathologic findings led to the diagnosis of hyaline vascu-



Fig. 6 Enhanced CT scan

A CT scan performed 8 months after surgery shows no recurrent tumor, while it reveals marked shrinkage of the retroperitoneal lymph nodes (arrows).

lar type CD. An early stage of the same histologic change was seen in the adjacent small lymph node.

The patient did not undergo radiation therapy or chemotherapy. A CT scan performed 8 months after surgery showed no recurrent tumor, and marked shrinkage of the retroperitoneal lymph nodes (Fig. 6).

### III. Discussion

Castleman's disease, for which the etiology is still obscure, was first described as a pathologic entity in 1954<sup>2)</sup>, and was later defined by Castleman et al. in 1956<sup>3)</sup>. CD develops mainly in young adults, although the age range is quite wide (8 to 66 years). CD occurs equally in both males and females<sup>3)</sup>. Although the tumor may arise in any area of the body where lymphoid tissue is found<sup>4)</sup>, the majority of cases arise in the organs and tissues cephalad to the diaphragm. The incidence of the tumor arising in the retroperitoneal space is approximately 12% of CD cases<sup>1)</sup>.

The clinical findings can be divided into two types: the unicentric type (UC) and the multicentric type (MC)<sup>3)</sup>. The UC type is characterized by an asymptomatic and solitary mass, whereas the MC type is characterized by systemic symptoms, such as fever, anemia, weight loss, and other manifestations<sup>5)</sup>. The UC type of CD is rarely associated with systemic symptoms (<10%)<sup>6)</sup>, and none were reported in the case we present here.

The histologic findings can be divided into two types: the hyaline vascular (HV) and the plasma cell (PC) type<sup>3)</sup>. The PC type tumor, which accounts for less than 10% of CD, has a spreading arrangement of the plasma cells in the involved lymph nodes. The HV type tumor is present in more than 90% of CD cases, and is characterized by small hyaline follicles and intrafollicular capillary proliferation. The patients with the HV type tumor are usually asymptomatic, as was observed in the present case, whereas those with the PC type may have systemic manifestations and abnormal blood tests, including an elevated ESR, hypergammaglobulinemia, and an increased level of C-reactive protein. The changes observed in patients with the PC type of tumor are caused mainly by the overproduction of IL-6 by the tumor<sup>6)</sup>. In the present case, although the serum level of IL-6 was not elevated, ESR was elevated and the retroperitoneal tissue was involved in severe chronic inflammation. Therefore, the tumor in the present case had some of the biological characteristics of the PC type CD, although the tumor lacked the histologic features of the PC

type CD.

The incidence of the tumor arising in the retroperitoneal space is low (0.2% of all tumors), and the differential diagnosis presents considerable difficulty. Eighty to 85% of retroperitoneal tumors have a high malignant potential, and liposarcoma, leiomyosarcoma, and malignant histiocytoma comprise 60 to 80 % of this group<sup>7)</sup>. In the present case, the preoperative imaging findings differed from those of frequently occurring tumors, causing us to consider the possible diagnosis of CD<sup>8) 9) 10)</sup>. Inflammatory myofibroblastic tumor (IMT), also called inflammatory pseudotumor, is usually found in the lung. However, 5% of IMT in the extra pulmonary sites are also found in the retroperitoneal space<sup>11)</sup>. The histologic findings of IMT are proliferating myofibroblasts with an associated inflammatory component<sup>12)</sup>. In the present case, the histologic findings differed from those of IMT.

When the tumor is the localized type of CD, surgical resection of the tumor is frequently the most effective treatment, thus yielding a favorable outcome, as neither radiotherapy nor chemotherapy is effective<sup>7)</sup>. Because the patient in the present case had sparse symptoms and signs, and few abnormal blood tests, we considered the possibility that the tumor was a UC type CD, and that the swelling of the surrounding lymph nodes was caused by the response to the tumor presence.

Swollen lymph nodes shrunk after surgery, in this case, but an early stage of the same histologic change was seen in the adjacent small lymph node. Some researchers have reported swollen lymph nodes, at a distance from the tumor, have been the reactive lymphoid hyperplasia<sup>13) 14)</sup>. Murashita et al. reported that lymph node adjacent the tumor was similar to it histologically in HV type and UC type of CD<sup>15)</sup>, as we experienced. We therefore think that a resection of the lymph nodes adjacent the tumor should be performed in order avoid local recurrence.

Larroche et al. reported the concurrence of malignant lymphoma among patients with CD<sup>16)</sup>, therefore necessitating further follow up study.

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# 後腹膜腔に発生した, Unicentric type Castleman's disease の一例

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## 要 約

患者は44歳, 男性。定期検診で腹部腫瘤を指摘されたため, 検査, 治療目的で当院へ紹介された。症状はなく, 血液検査結果では赤沈の亢進を認めるのみであった。身体所見では左上腹部に腫瘤を認めた。術前の画像診断により, 後腹膜腔に発生した Unicentric type Castleman's disease を疑った。また, 大動脈周囲リンパ節

の腫脹を認めた。開腹術を施行したところ, 腫瘍は後腹膜腔に存在し, 慢性の炎症により周囲組織に強く癒着していたが, 摘出することができた。病理学的には Hyaline vascular type Castleman's disease と診断された。術後8ヶ月が経過したがあきらかな再発を認めず, CT上, 大動脈周囲のリンパ節は縮小傾向である。

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