

## Castleman's Disease of the Retroperitoneum

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**SUMMARY :** A case of retroperitoneal CASTLEMAN's disease of hyaline-vascular type is presented. A 58-year-old woman was admitted with a four month history of lower back pain. Although a retroperitoneal tumor was found on sonography and CASTLEMAN's disease was included in the differential diagnosis, further imaging procedures such as computed tomography and angiography could not confirm the preoperative diagnosis of CASTLEMAN's disease. After the surgical removal of the tumor, the patient has been doing well with no evidence of recurrent disease.

### INTRODUCTION

CASTLEMAN's disease is a rare benign lymphoid tumor of unknown etiology. Since CASTLEMAN, *et al*<sup>1)</sup> first described a group of patients with a large thymoma-like mass in the anterior mediastinum, hundreds of cases have been reported in the literature. The mediastinum is the most common location, accounting for two-thirds of the cases. The superficial form involves the cervical, axillar, or inguinal lymph nodes and accounts for about 20% of the reported cases. The abdominal location is uncommon and this group consists of intra-peritoneal and retroperitoneal tumors. Most of the retroperitoneal tumors are located in the pelvis, and the retroperitoneal tumors involving the pararenal area are rare<sup>2)</sup>. In this paper a case of retroperitoneal CASTLEMAN's tumor of hyalin-vascular type is reported.

### CASE REPORT

A 58-year-old Japanese woman developed

lower back pain of four month duration. She was diagnosed as having a retroperitoneal tumor on a sonographic examination of the abdomen. She was admitted to the Department of Radiology, Nagasaki University Hospital in September 1987 for further evaluations of the tumor. Although CASTLEMAN's disease was included at the top in the differential diagnosis, a definite preoperative diagnosis was not established. She was transferred to our care for extirpation of the tumor. Her past history included bronchial asthma requiring medication for years and an operation for uterine myoma at the age of 55. Her family history was not contributory.

A physical examination on admission revealed an obese and fit middle-aged woman with a previous operation scar along the lower midline of the abdomen. No tumor was palpated. Laboratory data showed no hematologic abnormalities, including a hemoglobin level of 13.6g%, negative C-reactive protein, normal erythrocyte sedimentation rate of 13mm/hour, lactate dehydrogenase of 228 IU/L, total protein of 6.6g% with r-globulin of 17.3%.

Alpha-fetoprotein, carcinoembryonic antigen, and carbohydrate antigen 19-9 were all within normal limits. Serum levels of adrenaline, noradrenaline, dopamine were normal. There were no immunologic abnormalities. The OK-T<sub>4</sub> was 28.6% (normal : 30-45%) and OK-T<sub>8</sub> 29.0% (normal : 25-35%).

Abdominal sonography showed a hypoechoic sonolucent mass in the left paraortic region, measuring 3 cm by the greatest dimension (Fig. 1). Computed tomography (CT) revealed a well-demarcated homogenous mass of soft tissue density (Fig. 2), which was promptly and remarkably enhanced by contrast media. The abdominal aortic angiograms demonstrated the hypervascular tumor located between the left kidney and aorta. The left renal arterio-



Fig. 1. Abdominal sonography showing a hypoechoic sonolucent mass in the left paraortic region, measuring 3 cm by the greatest dimension (arrows)

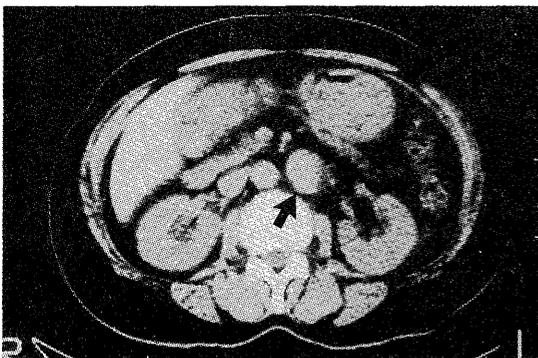


Fig. 2. Computed tomography (CT) revealing a well-demarcated homogenous mass of soft tissue density (arrow)

grams provided no further information. The second lumbar arteriograms showed a hypervascular mass (Fig. 3) with no findings of malignancy. Methyl iodo-benzyl guanidine (MIBG) scintigrams revealed no uptake by the tumor on the corresponding area.

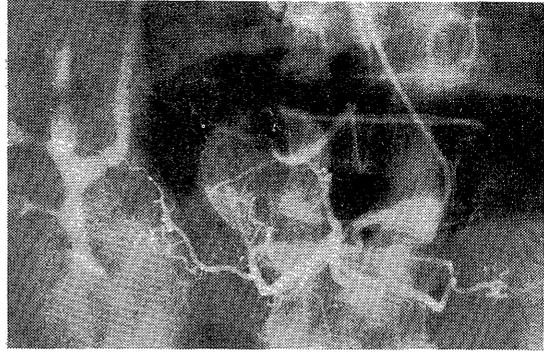


Fig. 3. Second lumbar arteriography showing a hypervascular mass with no malignant findings

The operation was performed on October 6, 1987. At laparotomy an elastic soft tumor surrounded by fat tissue was palpable between the aorta and left kidney. The left renal vessels were stretched upwards over the superior surface of the tumor. The tumor appeared to be mainly perfused by branches of the lumbar artery and drained via two branches into the left renal vein. The tumor was well-delineated and there were no findings of tumor invasion to the adjacent organs and structure. The tumor was extirpated together with the surrounding fat and two lymph nodes of the adjacent paraortic area. A frozen section during operation established the diagnosis of CASTLEMAN'S disease with no evidence for malignancy.

The resected tumor weighed 37 grams. Its cut surface showed a well-defined capsule and a homogeneous, greyish-white, fleshy texture with some scattered small bleeding spots. Pathologic features on a permanent section confirmed a hyaline-vascular type of benign giant lymph node hyperplasia or CASTLEMAN'S disease. The lymph nodes obtained from the paraaortic area showed normal findings.

Her postoperative convalescence was uneventful. At follow-up 1.5 years later the patient is well with no symptoms or signs of recurrent

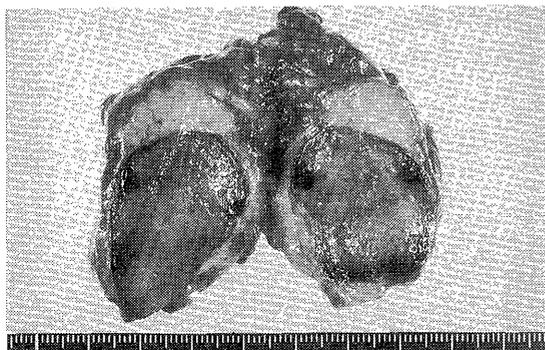


Fig. 4. The cut surface of the resected tumor showing a well-defined capsule and a homogeneous, greyish-white and fleshy texture with some scattered small bleeding spots

disease on abdominal computed tomography (CT) and sonography as well as clinically.

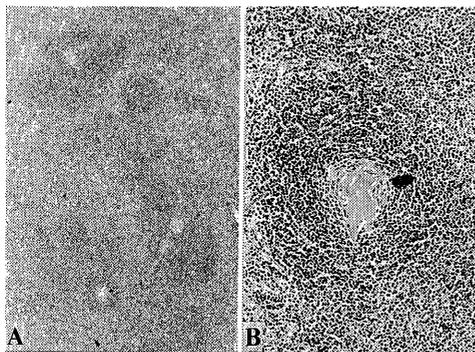


Fig. 5. Pathological findings of the tumor showing a hyaline-vascular type of benign giant lymph node hyperplasia or Castleman's disease. A : hematoxylin-eosin stain  $\times 10$ , B : hematoxylin eosin stain  $\times 50$

## DISCUSSION

The etiology of CASTLEMAN'S disease still remains controversial. Some authors consider it an inflammatory process in origin and others suggest a hamartomatous origin<sup>3)</sup>. Some reports show markedly increased anti-Ebstein Barr (EB) virus antibody<sup>4, 5)</sup> suggesting that EB virus be related to the etiology of CASTLEMAN'S disease.

The lesions are discovered most often either on routine roentgenograms of the chest or

because of pressure symptoms. The lesions sometimes manifest as palpable mass if outside the thorax. On sonography and CT scan the tumor manifests multinodular pattern. Angiograms show enlarged feeding vessels, dense homogeneous tumor stain, irregular hypervascularity, corresponding well to the prominent vasculature seen on histologic features<sup>6, 7)</sup>.

KELLER and associates<sup>8)</sup> undertook a clinicopathological analysis of 81 cases of CASTLEMAN'S disease and identified two histopathologic subtypes : the hyaline-vascular type and plasma cell type. These subtypes of the disease reflect quite different clinical manifestations. The hyaline-vascular lesions are most numerous accounting for 91% of cases, and are characterized by small hyalinized vascular follicles and interfollicular capillary proliferation. The plasma cell type, representing the remainder, are characterized by large follicles with intervening sheets of plasma cells<sup>8)</sup>. FLENDRIG<sup>9)</sup> observed an intermediate group with the characteristics of large follicles with intervening sheets of plasma cells<sup>8)</sup>. FLENDRIG<sup>9)</sup> observed an intermediate group with the characteristics of the two types, which he called the transitional form. Approximately 70% of cases of hyaline type are present as a solitary mass, whereas multiple are predominant in plasma cell type (about 60%)<sup>10)</sup>. The plasma cell type is often associated with systemic manifestations. A number of unusual clinical entities have been associated with the plasma cell type of classical CASTLEMAN'S disease, including fever, anemia, hyperglobulinemia, amyloidosis, nephrotic syndrome, and thrombocytopenia. All of those have at least partially responded to surgical removal of the affected lymphoid mass. Extirpation of the tumor is, therefore, the treatment of choice. Local recurrence after operation does not occur. When symptoms persist, a diagnosis of partial removal of the lesion or of a multicentric type of the disease is indicated. If complete removal is not possible, even partial excision may be useful since regrowth of the tumor is not expected. Only a very small number of cases, however, evolve into malignant lymphoma<sup>11)</sup>. Radiotherapy has produced little or no shrinkage of CASTLEMAN'S tumors<sup>8)</sup>.

The recent literature, however, has described a highly lethal "multicentric" variant of classical CASTLEMAN'S disease with similar hyperplastic angio-follicular morphologic features and a positive human immunodeficiency virus (HIV), suggesting that HIV may be a potential etiopathogenetic agent on patients with generalized multicentric CASTLEMAN'S disease<sup>12)</sup>.

With the advance in several non-invasive modalities for diagnosis, it is anticipated that incidental detection of asymptomatic cases of CASTLEMAN'S disease will be increased.

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