

# Castleman Disease in the Kidney and Retroperitoneum Mimicking Renal Cell Carcinoma with Retroperitoneal Lymphadenopathy: A Case Report\*

국소 림프절 종대를 동반한 신세포암처럼 보였던 신장과 후복막강을 침범한 캐슬만병: 증례 보고

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Castleman disease, or angiofollicular lymph node hyperplasia, is a fairly rare benign tumor of lymphoid origin with unknown etiology. Castleman disease arises mostly in the mediastinum, and some cases of renal and retroperitoneal involvement have been reported. However, Castleman disease that simultaneously involves the kidney and regional lymph nodes has not been reported in radiologic literature. We report a case of renal and pararenal Castleman disease, mimicking renal cell carcinoma with retroperitoneal lymphadenopathy.

## Index terms

Castleman Disease  
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CT

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## INTRODUCTION

Castleman disease (CD) is an uncommon benign lymphoproliferative disorder, characterized by hyperplasia of lymphoid follicles. Histologically, CD can be classified into hyaline-vascular and plasma cell types. And clinically, it also can be divided into localized and disseminated types. Ninety six percent of localized CD is hyaline vascular type, and on the other hand, disseminated CD is usually a plasma cell type. CD is commonly located in the mediastinum. Retroperitoneal and pararenal locations are relatively uncommon accounting for 7% and 2% of all cases, respectively. To date, to the best of our knowledge, there have been 100 cases of renal involvement in CD. Sixty eight cases were disseminated and 32 cases were localized CD. Among localized CD,

only 6 cases confined to the kidney have been reported (1-8). There are no reports in the radiologic literature describing CD, involving both kidney and pararenal space, mimicking renal cell carcinoma with lymph node metastasis. Here, we present a case of renal and pararenal CD misdiagnosed as a neoplasm of the kidney with retroperitoneal lymphadenopathy.

## CASE REPORT

A 36-year-old man was hospitalized in our institute, complaining of abdominal discomfort. The physical examination showed no remarkable finding, such as a palpable mass or tenderness. Complete blood count and routine blood biochemistry were within normal limits and the results of urinalysis re-

vealed no evidence of hematuria. The abdomen CT presented a mass with  $2.4 \times 2.1$  cm in size that was well demarcated in the upper pole of the left kidney. On a noncontrast CT scan, the mass appeared iso- to slightly high attenuating relative to the kidney parenchyma (Fig. 1A). In corticomedullary phase, the tumor enhanced homogeneously, with slightly lower attenuation than that of the normal renal parenchyma, measuring 109 HU (Fig. 1B). In the nephrographic phase, the tumor measures 95 HU, which was only slightly lower than in the corticomedullary phase (Fig. 1C). Another noteworthy finding was that there were several enlarged lymph nodes in the left pararenal space. All the lymph nodes showed the same enhancement pattern as the renal mass, with Hounsfield unit of 95-99 and 88-90 in corticomedullary and nephrographic phase, respectively (Fig. 1A-C). Moderate homogeneous enhancement of the renal mass and regional lymphadenopathy suggested the diagnosis of the primary renal neoplasm, such as renal cell carcinoma; although some features were not typical for conventional renal cell carcinoma. The renal mass and lymph nodes demonstrated a slow washing out pattern, and did not show marked Hounsfield unit difference between corticomedullary and nephrographic phase. In addition, the mass was not expansile nor exophytic, and pseudocapsulation of the mass was not definite. However, these findings are nonspecific and remain suspicious for renal neoplasm.

Patient underwent partial nephrectomy and regional lymphadenectomy. Grossly, the tumor specimen showed a well demar-

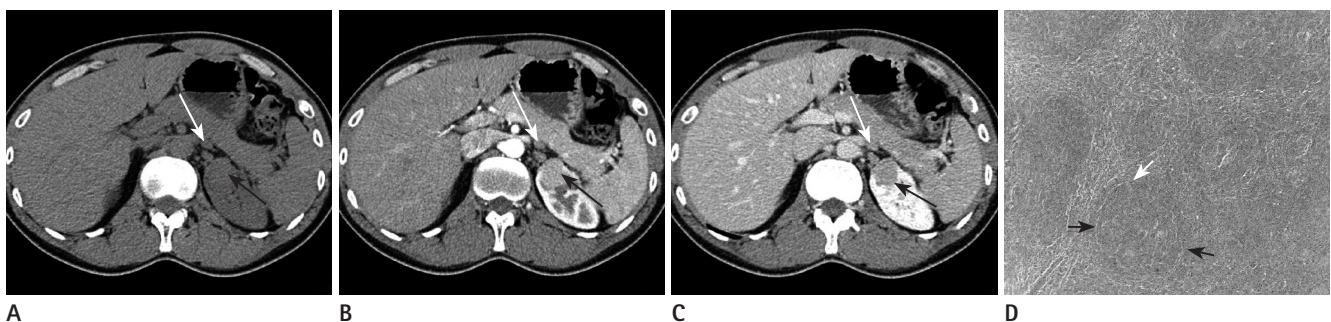
cated yellowish mass, measuring  $2.2 \times 1.8$  cm in size. Pathologic examination of the resected tumor from the left kidney and retroperitoneal lymph nodes showed increased lymphoid follicles with relatively small germinal centers. These germinal centers were penetrated by hyalinized venules and the follicles were separated by hypervascular interfollicular tissues. These findings were consistent with a diagnosis of hyaline vascular type CD (Fig. 1D). We also performed immunohistochemical study to rule out B cell lymphoma. On immunohistochemical staining, CD 20 was positive, but CD 5, 10 and Bcl-2, 6 were negative, with which B-cell lymphoma can be excluded.

The patient had uneventful postoperative course and has experienced no local or metastatic recurrence for two years after the surgery.

## DISCUSSION

Castleman disease (CD), which is also called angiofollicular lymph node hyperplasia, giant lymph node hyperplasia, lymphoid hamartoma and follicular lymphoreticuloma, represents not a neoplasm, but a morphologically distinct form of a rare atypical lymphoproliferative disorder. According to the literature, the disease is mainly in the thoracic (70%), followed by the cervix (14%), retroperitoneum (7%), axillary region (4%) and pararenal location (2%) (1-8).

CD can be classified into two pathological subtypes; "Hyaline



**Fig. 1.** A 36-year-old man with Castleman disease involving both kidney and pararenal space.

**A.** Noncontrast CT of abdomen shows iso to slightly high attenuated mass (black arrow) in the upper pole of the left kidney. And, there are several enlarged lymph nodes (white arrow) at the left pararenal space.

**B.** Contrast enhanced CT in corticomedullary phase shows homogeneous enhancement of the tumor (black arrow). Note that the several enlarged lymph nodes (white arrow) at left pararenal space demonstrate similar density of enhancement as the mass in the kidney, with Hounsfield unit of 109 and 95-99 in renal mass and lymph nodes, respectively.

**C.** Contrast enhanced CT in nephrographic phase reveals slow wash out enhancement pattern of the left renal mass (black arrow) and retroperitoneal lymph nodes (white arrow), with Hounsfield unit of 99 and 88-90 in the renal mass and lymph nodes, respectively.

**D.** Microscopic finding demonstrates increased lymphoid follicles with small germinal centers (black arrows) traversed by hyalinized blood vessels (white arrow). These findings are characteristics of the Castleman disease (Hematoxylin-Eosin,  $\times 200$ ).

vascular type” and “Plasma cell type”. The hyaline vascular type accounts for approximately 90% of CD, and is usually a solitary tumor, characterized by giant lymphoid follicles with a hyalinized central vessel. If there are more plasma cells in the tumor, the disease can be classified as a plasma cell type (9). And clinically, CD may also be categorized into two subgroups, according to the extent of the spread; localized and disseminated type. The two clinical types have distinct pathological and biological differences. Of the localized CD, 96% is hyaline vascular type with favorable prognosis after excision of the tumors. The pathology of disseminated CD is mainly a plasma cell type, which is treated with chemoradiation, but the prognosis is poor (10).

Localized CD usually manifests on CT as a single, well demarcated, and homogeneous mass with soft-tissue attenuation. The enhancement pattern of hyaline vascular CD is characterized by homogeneous enhancement in the early phase and persistent enhancement in the delayed phase. Relatively strong enhancement can be attributed to the abundance of blood supply and focal vascular proliferation of capillary vessels. Another distinguishing radiologic finding of localized CD is the absence or rare presence of cystic or necrotic degeneration in the tumor. This might be due to good circulation and low susceptibility of lymphatic follicles to necrosis (10). Additionally, Kim et al. (9) reported another unique feature of localized hyaline vascular CD of the abdomen; a single dominant mass with small satellite nodules, which is suggestive of regional lymphadenopathy. A main mass was found in various locations, including perirenal, para-aortic, mesenteric, perigastric, peripancreatic and adrenal regions, but not in the kidney.

On the other hand, plasma or disseminated type CD is characterized by the infiltration of plasma cells in and between the lymphoid follicles, and vascular proliferation is rare. Therefore, a strong enhancement of the lesion is not definite in this type (10).

The radiologic differential diagnosis of CD in the kidney and pararenal space would be lymphoma, angiomyolipoma with minimal fat, accessory spleen, and most importantly, renal cell carcinoma. Conventional renal cell carcinoma originates from the renal cortex, and typically exhibits an expansile growth pattern. Strong enhancement and rapid washing out pattern may help distinguish conventional renal cell carcinoma from other malignancies (11). Homogeneity of tumor enhancement are valuable CT finding for lymphoma, and angiomyolipoma with

minimal fat can be another differential diagnosis when the mass is high attenuating on noncontrast CT and shows homogeneous, prolonged enhancement pattern. However, all of these imaging findings are overlapped, which make accurate diagnosis of enhancing renal mass very challenging.

Complete surgical resection in an en-bloc manner, if possible, is nearly always successful, regardless of the type. The surgical removal of involved lymph nodes can resolve local and systemic symptoms and is associated with a good prognosis (12).

In conclusion, due to the overlapping imaging characteristics of renal cell carcinoma, lymphoma, angiomyolipoma with minimal fat and CD, preoperative diagnosis of CD is difficult. Therefore, surgical resection and histological evaluation should be mandatory to make an accurate diagnosis. However, if there is a strong enhancing renal mass with regional lymphadenopathy, CD can be included in the differential diagnosis, in addition to the primary and secondary renal neoplasms.

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## 국소 림프절 종대를 동반한 신세포암처럼 보였던 신장과 후복막강을 침범한 캐슬만병: 증례 보고

고희선 · 우지영 · 홍혜숙 · 정아영 · 양 익 · 이 열

캐슬만병은 매우 드문 림프구양의 증식성 질환으로, 대개는 종격동에서 발생하나 신장이나 후복막강에서 발생한 경우도 여러 차례 보고된 바 있다. 하지만 신장과 신장 주위 공간을 동시에 침범하여 국소 림프절 종대를 동반한 신장암과 유사한 모습을 보였던 경우는 매우 드물고, 그 영상의학적 소견의 보고는 아직까지 없어 증례 보고를 하고자 한다.

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