

Adrenal Castleman's Disease Mimicking Other Adrenal Neoplasms: A Case Report

부신의 다른 신생물을 오인할 수 있는 부신의 캐슬만병: 증례 보고

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We present a rare case of adrenal Castleman's disease with hyaline vascular type mimicking other adrenal neoplasms in a 65-year-old woman. Although rare, the hyaline vascular type of adrenal Castleman's disease should be included in the differential diagnosis if an adrenal mass shows a well-defined, highly enhancing solid adrenal mass with peripheral rim enhancement, multiple satellite lymph nodes, and peritoneal thickening around the dominant mass on computed tomography as shown in this patient.

Index terms

Giant Lymph Node Hyperplasia

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INTRODUCTION

Castleman's disease, also known as angiofollicular lymph node hyperplasia or giant lymphoid nodular hyperplasia, is a rare lymphoproliferative disorder. The mediastinum is the most common involvement site of Castleman's disease, although extrathoracic involvements including nodal and extranodal presentations have been described (1). Castleman's disease of the adrenal gland is very rare. We present a case of adrenal Castleman's disease mimicking adrenal neoplasm on multidetector computed tomography (CT).

CASE REPORT

A 65-year-old woman was referred to our hospital for abdominal discomfort of 2-weeks duration. Physical examination and routine laboratory test results were unremarkable. She had

a history of hypertension. Pre-enhanced CT scan revealed a well-defined homogenous soft tissue density mass (5.0 cm in long dimension) in the left adrenal gland with CT attenuation value of 41 Hounsfield unit (HU) (Fig. 1A). Contrast-enhanced CT scan showed a highly enhancing adrenal mass with CT attenuation value of 138 HU. Distinctive peripheral enhancement of the mass was also noted (Fig. 1B). There were multiple small enhancing retroperitoneal lymph nodes. Peritoneal thickening around the dominant mass was also noted (Fig. 1C). She had no history of malignancy. Therefore, our radiologic diagnosis was adrenal cortical carcinoma with metastatic lymph nodes. Differential diagnosis included malignant pheochromocytoma with metastatic lymph nodes.

The patient underwent a left adrenalectomy which revealed a hypervascular mass in the left adrenal area and several surrounding lymph nodes. During the operation, severe adhesions with peritoneal hyperplasia were noted.



Fig. 1. Adrenal Castleman's disease mimicking other adrenal neoplasms in a 65-year-old woman.

- A.** Pre-enhanced CT showing a well-defined homogenous soft tissue density mass of 5.0 cm in long dimension (arrow) in the left adrenal gland.
B. Contrast-enhanced CT showing a highly enhancing mass with peripheral rim enhancement (arrow). Peritoneal thickening (arrowhead) surrounding the mass is noted.
C. Contrast-enhanced CT showing multiple small enhancing retroperitoneal lymph nodes (arrow). Increased fat stranding in the left perirenal space was also noted.

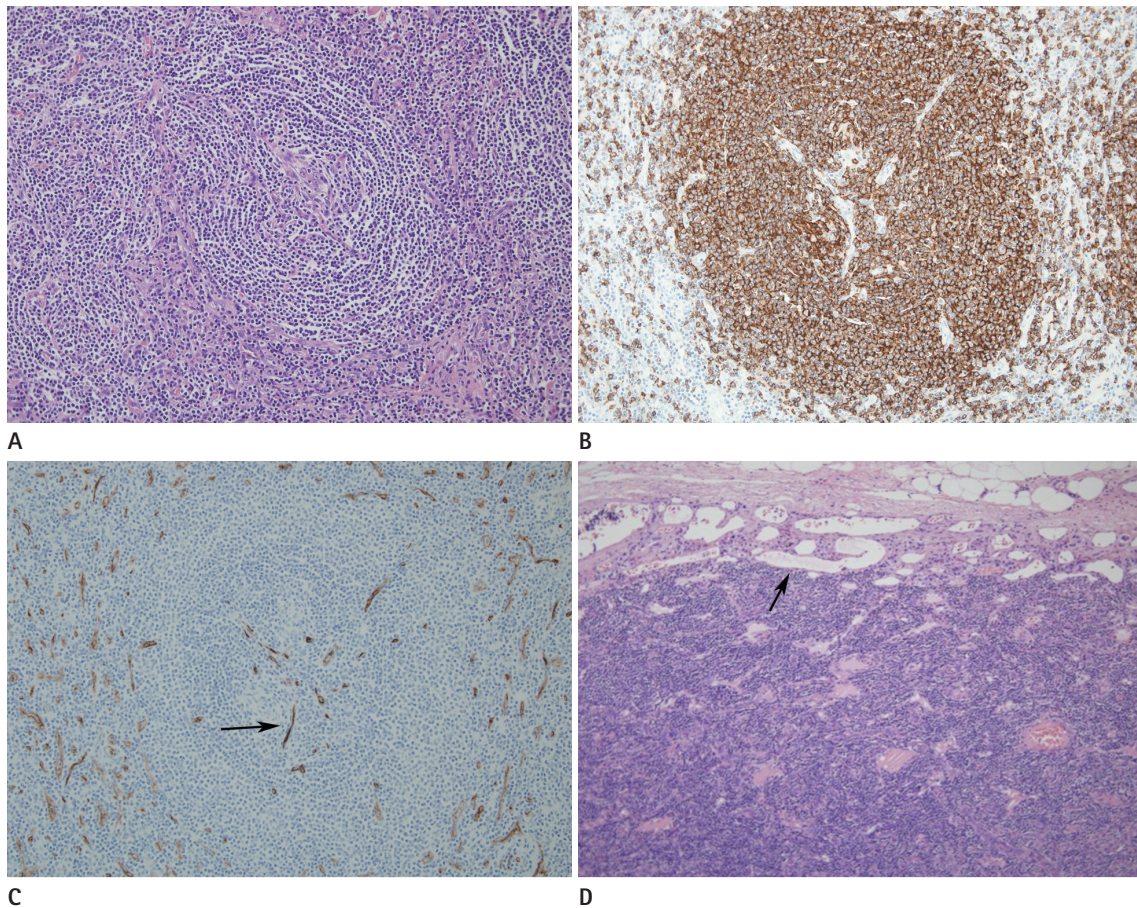


Fig. 2. Hyaline-vascular type Castleman's disease of the adrenal gland in a 65-year-old woman.

- A.** Photomicrograph (original magnification, $\times 200$ hematoxylin-eosin stain) of the mass showing lymphoid follicles with small hyalinized germinal centers and broad mantle zone. Mantle zone lymphocytes are arranged in concentric rings ("onion skin" pattern).
B. The "onion skin" pattern is highlighted by immunochemical staining for CD20 (original magnification, $\times 200$).
C. Photomicrograph (original magnification, $\times 200$; immunochemical staining for CD34) of the mass showing some follicles with penetration by blood vessels ("lollipop follicle", arrow).
D. Photomicrograph (original magnification, $\times 100$ hematoxylin-eosin stain) of the mass showing prominent vascular proliferation (arrow) in the periphery of the mass.

Grossly, a relatively well-defined pinkish round adrenal mass (3.6 cm in long diameter) was seen. Microscopically, the mass contained lymphoid follicles with small hyalinized germinal centers with a broad mantle zone. The mantle zone lymphocytes were arranged in concentric rings (in an “onion skin” pattern). They were highlighted by CD20 immunochemical staining (Fig. 2A, B). There were some follicles penetrated by blood vessels known as “lollipop follicle”. They were highlighted by CD34 immunohistochemical staining (Fig. 2C). Prominent vascular proliferation at the periphery of the mass was also seen on hematoxylin-eosin staining (Fig. 2D). Accordingly, a diagnosis of hyaline-vascular type Castleman’s disease of the adrenal gland was made.

DISCUSSION

Castleman’s disease is a benign lymphoproliferative disease that preserves the lymph node architecture. Although its etiology and pathogenesis are poorly understood, chronic low-grade inflammation, immunodeficiency, and autoimmunity have been implicated (2).

There are morphological and histopathogenic classification system for this disease (3). The morphologic classification includes unicentric form and multicentric form based on the extent of local lymph node involvement. The histopathogenic classification of Castleman’s disease includes hyaline vascular, plasma cell, and mixed-type.

The hyaline vascular type, the most common pattern, has been seen in about 90% of cases. This type is characterized by abnormal lymphoid follicles, numerous vessels, and wide fibrous septa. The plasma cell type is a less common histological pattern. It is characterized by the presence of sheets of mature plasma cells and a few vessels (4). Recently, the plasma cell type is subdivided into two entities based on the presence or absence of human herpes virus-8 (2, 3).

The unicentric form is the hyaline vascular type. It occurs in 70–90% of cases. This type usually occurs in young adults. It is asymptomatic. The multicentric form occurs in older individuals. Approximately 80–90% of cases are plasma cell types. The multicentric form has a worse prognosis with systemic symptoms and signs compared to the unicentric form (3).

Castleman’s disease commonly involves the mediastinum.

However, it may occur anywhere along the lymphatic chain in the neck, axilla, thorax, abdomen, and pelvis. Extralymphatic sites of involvement include the lungs, larynx, parotid glands, pancreas, meninges, and muscles (5). Castleman’s disease of the adrenal gland is rare. Only a few cases have been reported (6). Moreover, imaging features of adrenal Castleman’s disease have not been well described.

The characteristic CT appearance of hyaline vascular Castleman’s disease is a solitary enlarged nodal mass or a dominant mass with surrounding small satellite nodules and intense enhancement (5). The intense enhancement is attributed to abundant blood vessels in the hyaline vascular type. The enhancement is generally homogenous, although heterogeneous enhancement is possible in larger lesions due to central necrosis. Approximately 10% of hyaline vascular Castleman’s disease has internal calcifications. They are characteristically coarse or with a distinctive arborizing pattern. Plasma cell Castleman’s disease typically demonstrates less avid enhancement compared to hyaline vascular Castleman’s disease.

In our case, the CT image showed a well-defined highly enhancing solid mass with homogenous soft tissue density. It was consistent with the imaging features of hyaline vascular Castleman’s disease in previous reports (7, 8). No distinctive arborizing calcification was seen in our case.

Recently, Zheng et al. (9) have reported new CT features of localized retroperitoneal Castleman’s disease, including peripheral rim enhancement and peritoneal thickening surrounding the mass. This literature explained that peripheral rim enhancement was attributed to the predominant peripheral small or capillary vessel on the mass while the peritoneal thickening around mass was attributed to the reactive peritoneal hyperplasia (9).

Our CT image also showed peripheral rim enhancement of the mass and surrounding peritoneal thickening. The peripheral enhancement of the mass on CT was correlated with marked vascular proliferation in the periphery of the mass microscopically. The surrounding peritoneal thickening on CT was correlated with severe adhesions and peritoneal hyperplasia around the left adrenal area seen at surgery. However, further studies with more cases are needed to determine whether these newly discovered CT features are specific for Castleman’s disease.

Castleman’s disease of the adrenal gland is so rare that its preoperative diagnosis is very difficult. It can mimic other hyper-

vascular adrenal tumors on CT. The differential diagnosis of adrenal Castleman's disease may include adrenocortical carcinoma and pheochromocytoma (10). An adrenocortical carcinoma is typically a large mass (the majority is measured at more than 6 cm) and heterogeneous enhancement due to necrosis. The large size and heterogeneity are more reliable indicators for the diagnosis of adrenocortical carcinoma than washout values. In 19–33% of cases, calcifications have been identified, more commonly microcalcifications. Invasion of the inferior vena cava is also commonly seen in adrenocortical carcinoma (10). Pheochromocytoma shows variable imaging features. It is usually homogeneous and solid when small. However, larger lesions may appear as fatty, cystic, or calcified heterogeneous masses. Classically, pheochromocytoma is characterized as a brightly enhancing mass with a wide range in the degree of enhancement at CT (10).

Although adrenocortical carcinoma or pheochromocytoma usually shows heterogeneous attenuation on CT, most Castleman's disease shows homogeneous attenuation. Arborizing calcification is unique to Castleman's disease. However, due to the overlap of some radiologic findings and its rarity in the adrenal gland, pathologic examination is needed to diagnose adrenal Castleman's disease.

In conclusion, we present a rare case of adrenal Castleman's disease with the hyaline vascular type mimicking other adrenal neoplasms. Although rare, we suggest that the hyaline vascular type of adrenal Castleman's disease should be included in the differential diagnosis, especially if it is a well-defined, highly enhancing solid adrenal mass with peripheral rim enhancement, multiple satellite lymph nodes, and peritoneal thickening around the dominant mass.

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부신의 다른 신생물을 오인할 수 있는 부신의 캐슬만병: 증례 보고

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저자들은 65세 여성에서 부신의 다른 신생물을 오인할 수 있는 초자질 혈관형 캐슬만병에 관한 드문 증례를 보고한다. 부신에 초자질 혈관형 캐슬만병이 나타나는 것은 매우 드물지만, 컴퓨터단층촬영상 경계가 좋고, 조영 증강이 잘되는 부신의 종괴가 종괴 변연부의 조영 증강, 여러 개의 위성 림프절, 종괴 주변의 복막 비후 소견을 동반한다면, 부신의 캐슬만병도 감별 진단에 포함시켜야 할 것이다.

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