



# Hyaline-vascular Adrenal Castleman's Disease Mimicking an Adrenal Neoplasm

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## Abstract

Castleman's disease (CD), also known as angiofollicular lymph node hyperplasia, is an uncommon, benign B-cell lymphoproliferative disorder. Adrenal CD is an extremely rare lesion of the adrenal gland that mimics other adrenal neoplasms reported only in a few cases in the literature. We presented a large-sized right adrenal CD, which was found incidentally in a 46-year-old male patient during the evaluation for coronavirus disease-2019 pneumonia. Although imaging features overlap with other hypervascular adrenal neoplasms, CD should also be considered in the differential diagnosis of the adrenal masses, particularly when it is relatively well defined with homogenous attenuation and enhancement despite its large size.

**Keywords:** Castleman's disease, adrenal neoplasm, hypervascular adrenal mass

## INTRODUCTION

Castleman's disease (CD), also known as angiofollicular lymph node hyperplasia, is an uncommon, benign B-cell lymphoproliferative disorder that preserves the lymph node architecture. The mediastinum is the most common part of involvement, however anywhere in the body can be involved, including both nodal, and extranodal sites (1). These lesions are rarely reported in the literature as a mimicked of other neoplasms of the associated organs, including a mimicked of a gastric submucosal leiomyoma, a mass of pancreatic head, a mesenteric mass, or a mimicked of renal cell carcinoma (2-4). We present a large-sized right adrenal CD which was found incidentally in a 46-year-old male patient during the evaluation for coronavirus disease 2019 (COVID-19) pneumonia.

## CASE PRESENTATION

A 46-year-old male patient with a 25 pack-year smoking history was admitted to our hospital with suspicion of COVID-19. On the upper abdominal images of the chest computed tomography

(CT), a 56x30 mm sized relatively well-defined soft tissue density lesion compressing the inferior vena cava was incidentally established at the right adrenal location. Only the medial limb of the adrenal gland was visible and the interface of mass with the remaining parts of the adrenal gland could not be discerned (Figure 1). The density of the mass on non-contrast CT was incompatible with a lipid-rich adenoma. On wash-out imaging, progressive enhancement was observed as opposed to that expected from lipid-poor adenoma and pheochromocytoma. There was no associated retroperitoneal lymph nodes or peritoneal thickening around the mass. Magnetic resonance imaging (MRI) did not show the characteristic bright T2w signal intensity of the pheochromocytoma. On chemical shift imaging, there was no signal drop in the out of phase images to suggest intravoxel (intracellular) lipid (Figure 2). He had no history of malignancy. At that time, positron emission tomography-CT reported minimally increased fluorodeoxyglucose (FDG) uptake (maximum standardized uptake value: 3.5) (Figure 3). With the suspicion of malignancy, surgical excision with inevitable right adrenalectomy was performed. Histopathological evaluation of



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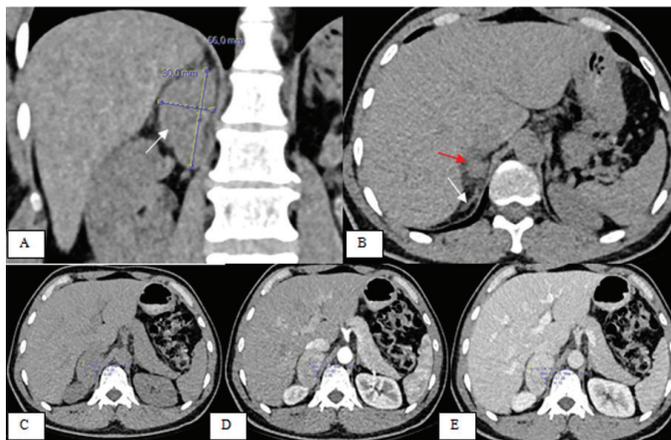
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the lesion revealed hyalin vascular type adrenal CD (Figure 4). Informed consent has been taken from the patient.

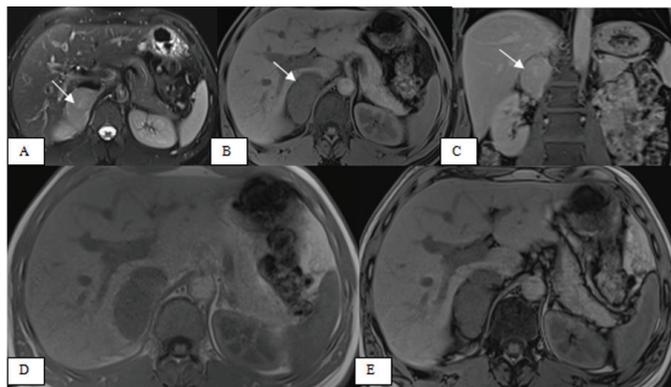
## DISCUSSION

CD is a rare benign lymphoproliferative disease with not well-known etiology and pathogenesis, although chronic low-grade inflammatory states and autoimmunity have been suggested (5). There are two distinct clinical subtypes including the unicentric (localized) CD and multicentric CD. Unicentric CD is more common occurring usually in young adults and is usually asymptomatic, if there are no symptoms resulting from the



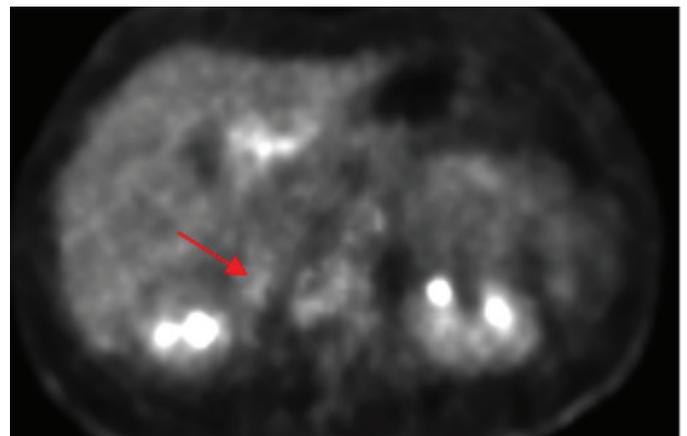
**Figure 1.** (A, B) Non-contrast CT images showing the right adrenal mass-measuring 56x30 mm on coronal image (A, arrow). On axial images the lateral limb of the right adrenal gland is shown, but corpus of the rest of the gland was not visible (B, white arrow). The part of the mass that should not be discerned from the adrenal gland is shown (B, red arrow). (C-E) Contrast washout CT scan demonstrates soft tissue density of the lesion on non-contrast CT (C) and progressive enhancement during the arterial (D) and venous (E) phases

CT: Computed tomography



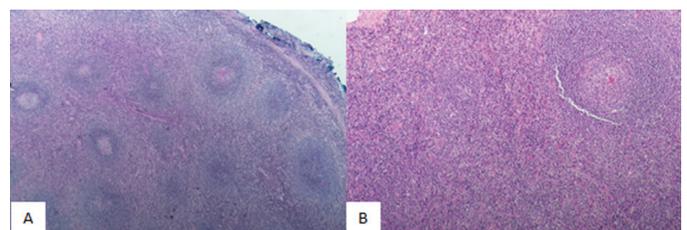
**Figure 2.** Magnetic resonance imaging showing the mass on T2w image having high signal intensity relative to the liver but not as much as expected from a pheochromocytoma (A, arrow). Precontrast T1w (B, arrow) and postcontrast T1w (C) images show homogenous enhancement of the lesion (C, arrow). There was no drop in signal intensity out of phase imaging (A; in phase and B; out of phase)

localized pressure of the mass. In our patient, the right adrenal mass was incidentally found during the evaluation of COVID-19 pneumonia at the upper abdominal images included to the chest CT. On the other hand, the multicentric CD, which occurs more commonly in older patients is a systemic disease associated with a poor prognosis (6,7). The histopathological variants are hyaline vascular type, plasma cell type, and mixed-type. While the unicentric CD occurs in the form of a hyaline vascular type, which is seen in about 90% of the cases, most of the multicentric CD demonstrates the plasma cell type (5,7). Reported in only a few cases in the literature, adrenal CD is an extremely rare mass lesion of adrenal gland mimicking other adrenal neoplasms (8,9). The hyaline vascular type is characterized with a high number of blood vessels in relation with abnormal lymphoid follicles in contrast to the plasma cell type, which is associated with little number of vessels along with mature plasma cells (4). Because to the rarity of the adrenal CD and the absence of well defined specific radiological imaging findings, the preoperative diagnosis is very difficult. On radiological imaging, the unicentric hyalin vascular type CD is seen as a solitary mass usually homogenous in attenuation with prominent and usually homogenous contrast enhancement due to its highly



**Figure 3.** FDG-PET/CT showing minimally increased focal FDG uptake at the central part of the lesion (arrow)

FDG: Fluorodeoxyglucose, PET: Positron emission tomography, CT: Computed tomography



**Figure 4:** Microscopic appearance of the lesion (A) hematoxylin-eosin stain X 40 and (B) hematoxylin-eosin stain X 100 show lymphoid follicles with small hyalinized germinal centers and a broad mantle zone in association with prominent vascular proliferation in interfollicular areas

vascular nature. Although adrenal cortical carcinoma and pheochromocytoma generally have generally heterogeneous attenuation and enhancement pattern, large CD lesions with necrotic parts may also show heterogeneous enhancement and should be considered in the differential diagnosis. In some case reports, peripheral rim like enhancement has been shown in association with marked peripheral capillary vessels seen on the microscopic evaluation of hyalin vascular CD. In these case reports, peritoneal thickening surrounding the mass has also been demonstrated and attributed to reactive peritoneal hyperplasia (9,10). Some internal calcifications may also present in the hyalin vascular CD. In our patient, the right large-adrenal mass showed soft tissue density on non-enhanced CT images without any peritoneal thickening and internal calcifications. Washout CT imaging and MRI have revealed progressive intense enhancement during arterial and venous phases. Additionally, the density on early phases was not as high as was expected from a pheochromocytoma. The absence of intracellular lipid has also been verified by chemical shift MRI with a demonstration of the absence of a signal drop on the opposite phase. Minimal FDG uptake was present. Although there were no areas of hemorrhage or necrosis as expected from such a large adrenal cortical carcinoma or there was no history of primary malignancy to suggest metastasis, due to the overlapping imaging findings, histopathological diagnosis was needed.

## CONCLUSION

Adrenal CD is an extremely rare lesions of the adrenal gland. Although imaging features overlap with other adrenal neoplasms, CD should also be considered in the differential diagnosis of the adrenal masses, particularly when it is relatively well defined with homogenous attenuation and enhancement despite its large size.

## Ethics

**Informed Consent:** Informed consent has been taken from the patient.

**Peer-review:** Externally peer-reviewed.

## Authorship Contributions

Concept: B.E., S.D.Ö., D.A., H.Ö., Design: B.E., S.D.Ö., D.A., Data Collection or Processing: B.E., T.O.K., İ.M., Analysis or Interpretation: B.E., H.Ö., Literature Search: B.E., T.O.K., Writing: B.E., İ.M.

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