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Practical uroradiology

Castleman's disease

Jia-Hwia Wang [a, b, c, *]

[a] Department of Radiology, Cheng Hsin General Hospital, Taipei, Taiwan, ROC
[b] Department of Radiology, Taipei Veterans General Hospital, Taipei, Taiwan, ROC
[c] School of Medicine, National Yang-Ming University, Taipei, Taiwan, ROC

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1. Introduction

Castleman's disease (CD) is a lymphoproliferative disease. Two histological subtypes of CD are hyaline-vascular and plasma cell variants. CD is classified clinicoradiologically into unicentric (localized) type and multicentric (diffuse) type according to the extent of spreading of the disease. Initial CD diagnosis is usually based on imaging findings; further confirmation is made by excisional biopsy and histopathological evaluation.

2. Case report

A 57-year-old female patient was admitted to hospital for further consultation regarding a left suprarenal tumor which was found incidentally during a physical check-up 6 years previously. This left suprarenal tumor showed an increase in size in a recent follow-up computed tomography (Fig. 1). On admission, physical and laboratory examinations were unremarkable. Following the initial examination, magnetic resonance imaging (MRI) revealed a left suprarenal tumor (Fig. 2). Robotic-assisted laparoscopic left-adrenalectomy and retroperitoneal lymph node dissection were performed due to the presence of a left-adrenal tumor with enlarged regional retroperitoneal lymph nodes. The pathological report revealed CD in the retroperitoneal lymph nodes, and normal left-adrenal gland. The postoperative course was uneventful. The patient was discharged in a stable condition. Outpatient follow up was recommended.

3. Discussion

CD was first reported by Castleman and Towne[1] in 1954. The cause of CD is still unknown. Two histological subtypes of CD are hyaline-vascular and plasma cell varaints.[2] CD is classified clinicoradiologically into unicentric (localized) type and multicentric (diffuse) type according to the extent of spreading of the disease.[3,4] CD may occur in any site where lymph nodes exist. Approximately 70% of CD occurs in the thorax (10–15% each in the neck and abdomen), retroperitoneum, and pelvis.[2,5] The hyaline-vascular variant accounts for approximately 90% of unicentric CD and is characterized by lymphoid follicular hyperplasia and vascular proliferation in the interfollicular region. The plasma cell variant is seen in both unicentric and multicentric forms of CD, and comprises approximately 10% of unicentric CD. It is characterized by sheets of polyclonal plasma cells within the interfollicular zone and more variable vascular proliferation compared to the hyaline-vascular variant. Both HIV and HHV-8 have been associated with CD.[6] The majority of multicentric CD is associated with an immunodeficiency state, such as HIV and HHV-8 infection.[7] Unicentric CD is usually seen in patients <30 years of age. It tends to affect nodal groups, and causes symptoms and signs from lymphadenopathy. Multicentric CD is usually associated with HIV and HHV-8 co-infection, and hence it has systemic symptoms and signs such as loss of appetite, weight loss, night sweats, and fatigue.

Initial CD diagnosis is usually based on imaging findings, and further confirmation is made by excisional biopsy and histopathological evaluation. CT generally shows a well-circumscribed mass of soft tissue attenuation. Calcification is infrequent and can include punctate, coarse, peripheral, and arborizing patterns. Smaller masses show homogeneous contrast enhancement, and larger masses show heterogeneous contrast enhancement.[8,9] Differential diagnosis varies according to the location such as paragangliomas, pheochromocytomas, neurogenic tumors, sarcomas, and

* Department of Radiology, Cheng Hsin General Hospital, Number 45, Cheng Hsin Street, Beitou, Taipei, 112, Taiwan, ROC.
E-mail address: wangjh@vghtpe.gov.tw.

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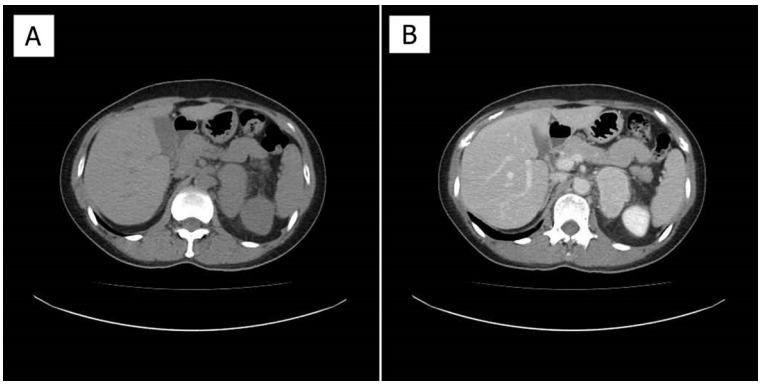

Fig. 1. Retroperitoneal Castleman's disease. (A) Axial noncontrast computed tomography reveals a well-defined mass with density identical to the muscle in the left suprarenal region. (B) Axial contrast-enhanced computed tomography reveals marked contrast enhancement of this mass.

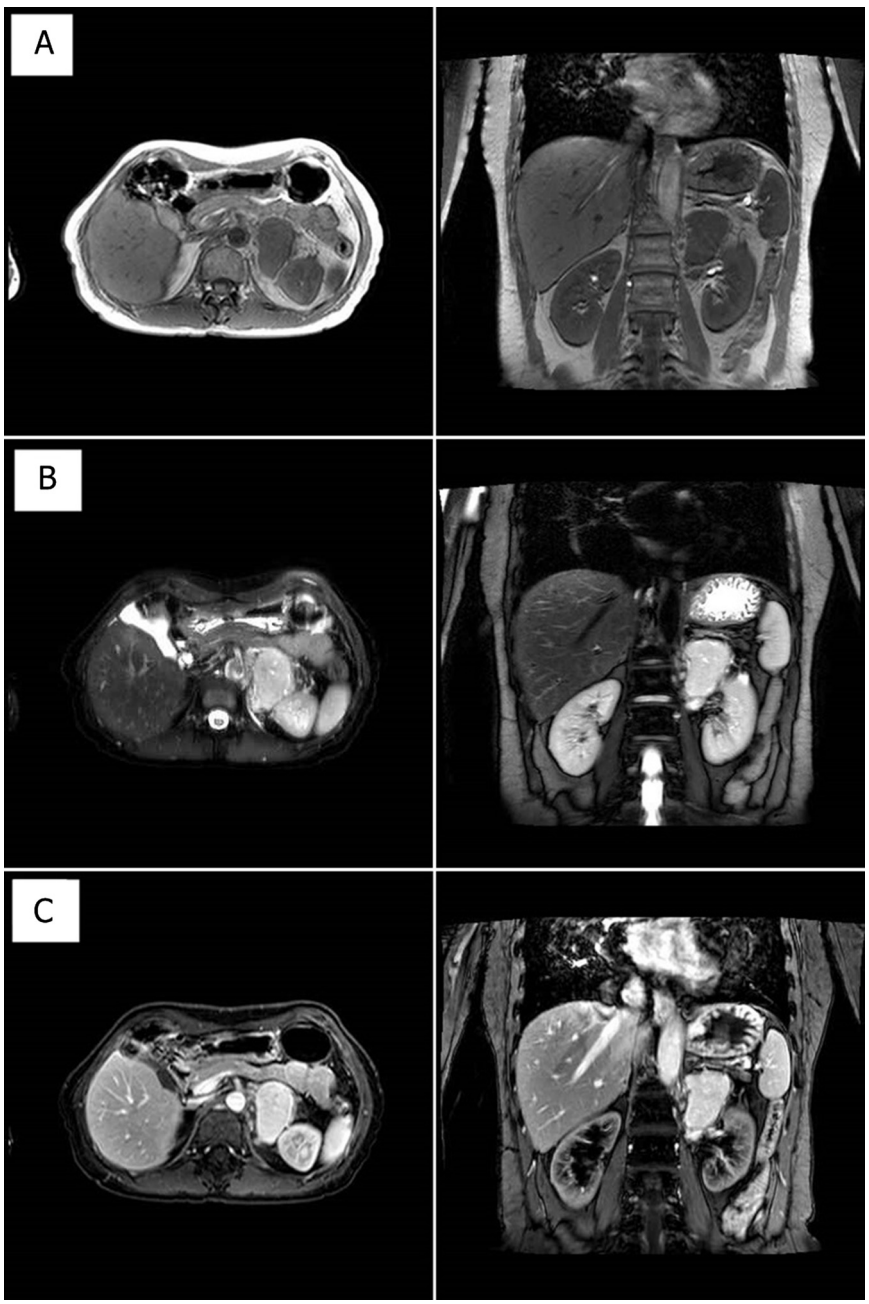

Fig. 2. Retroperitoneal Castleman's disease. (A) Axial and coronal noncontrast T1-weighted magnetic resonance image (MRI) reveals a well-defined mass with signal intensity identical to the muscle in the left suprarenal region. (B) Axial and coronal noncontrast T2-weighted MRI reveals high signal intensity of this mass. (C) Axial and coronal contrast-enhanced T1-weighted MRI reveals marked contrast enhancement of this mass.

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lymphomas. On MRI, CD shows identical or slightly high signal to muscle on T1-weighted images and hyperintense signals on T2-weighted images, and marked contrast enhancement.[10]

Treatment is surgical resection.

Conflicts of interest

The author declares no conflicts of interest.

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