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Case report

Castleman disease: a rare case in a young woman

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Abstract. Castleman disease is a rare lymphoproliferative disorder characterized by benign enlargement of lymph nodes. It is divided into unicentric disease, which involves a single enlarged lymph node, and multicentric disease, which affects multiple lymph node stations. In this report, we describe a rare case of a 28-year-old female patient with an unicentric Castleman disease. Computed tomography and magnetic resonance imaging revealed a well-circumscribed large mass in the left neck, characterized by intense homogenous enhancement and suspected for a malignant disease. The patient underwent an excisional biopsy for definitive diagnosis of unicentric Castleman disease and ruled out malignant conditions

Key words: Castleman disease (CD), lymph nodes, unicentric Castleman disease (UCD), excisional biopsy

Introduction

Castleman disease (CD) is a rare benign lymphoid disorder which presents more commonly as a localized solitary mass, known as the unicentric Castleman disease (UCD), whereas multicentric Castleman disease (MCD) is a systemic disease, characterized by generalized lymphadenopathy, constitutional symptoms, and more aggressive clinical course.

The incidence of UCD is 15 per million patient years (1) with no gender preference. The average age of diagnosis for UCD patients is typically 4th decade, younger than for MCD patients (6th decade) (2). The mediastinum is the most frequent location for UCD, with less common extrathoracic sites being the neck, axilla, abdomen, and pelvis, but can be found in any lymph node station. It usually has an indolent and non-progressive course in the vast majority of cases, but this condition has been associated with an increased risk of developing certain malignancies, as large B-cell lymphomas and follicular dendritic cell sarcomas (3).

Case report

A 28-year-old female patient presented to our institution with a large, left-sided, painless neck mass which had developed over 7 months. Physical examination was significant for a firm, nonulcerating lesion, with no overlying skin changes. Laboratory findings, including leukocyte and platelet counts, hemoglobin, serum creatinine, liver function did not reveal abnormalities. Contrast-enhanced computed tomography (CT) of the neck revealed a 28 × 56 × 20 mm well-circumscribed mass in the left neck characterized by intense homogenous enhancement (Figure 1). The lesion showed high signal compared to muscle on T1 and T2 weighted images in the subsequent magnetic resonance imaging (MRI) with intense *restricted* diffusion on diffusion-weighted imaging *(DWI)* and homogenous enhancement (Figure 2). Moreover, intralesional flow voids were seen on T1- and T2-weighted images reflecting the vascularity of the mass. A paraganglioma was firstly suspected although at an unusual site. The patient underwent

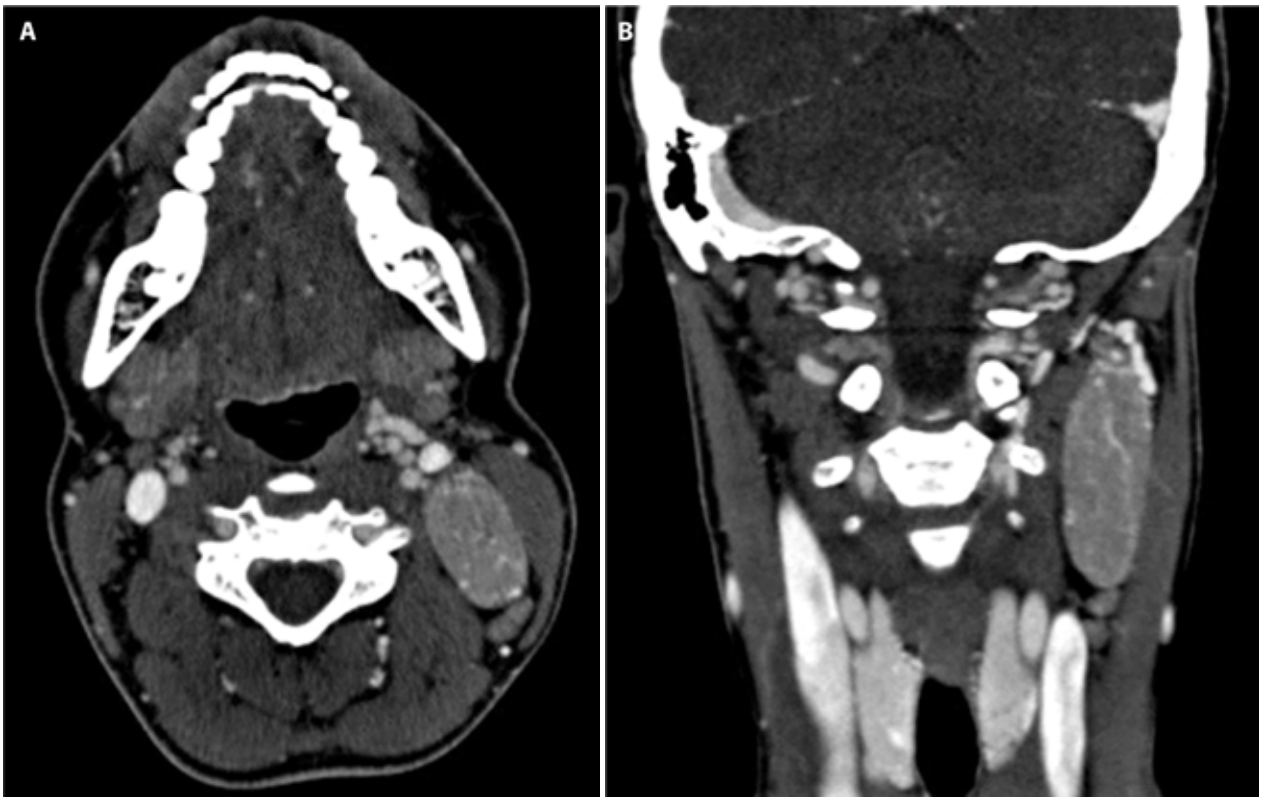

Figure 1. Contrast-enhanced axial (A) and coronal (B) neck CT scans which show a 28 × 56 × 20 mm well-circumscribed mass in the left neck characterized by intense homogenous enhancement.

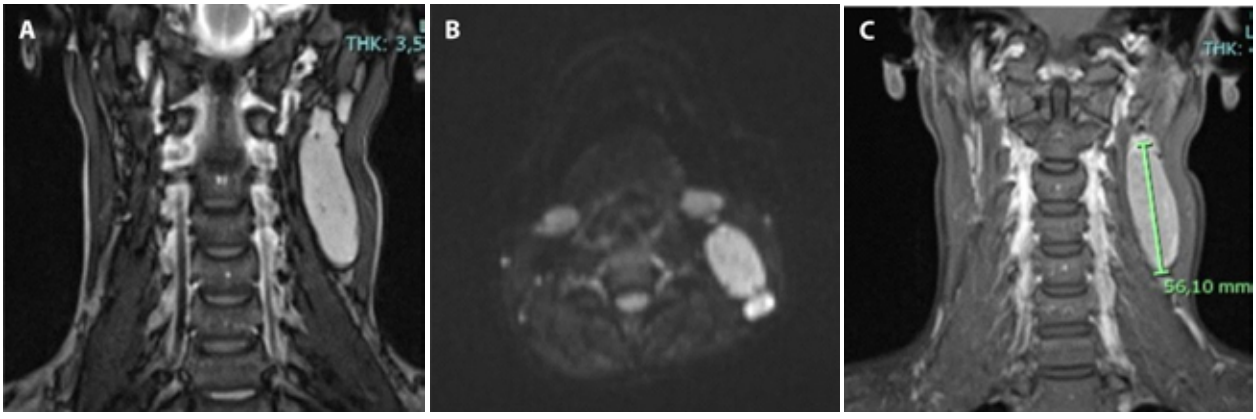

Figure 2. Coronal T2-weighted MRI image (A), axial DWI image (B) and coronal post-contrast T1-weighted MR image (C) demonstrating a hyperintense mass in the left neck, with intense *restricted* diffusion and homogenous enhancement.

an excisional biopsy for definitive diagnosis. Histopathologic examination revealed a lymph node with overall preserved architecture, containing numerous follicles of varying sizes with multiple hyalinized germinal centers. The mantle zones were expanded, with a concentric "onion skin" arrangement of lymphocytes. These findings were consistent with a diagnosis of UCD, hyaline vascular type. The patient currently remains stable with no signs or symptoms of recurrent disease for over 5 months.

Discussion

CD was first described in the 1950s by Benjamin Castleman as localized mediastinal lymph node enlargement characterized by a benign tumorous process of lymphocyte cell lines, whose multiplication leads to excessive expansion of lymph nodes (4). The mediastinum is the most common location for localized CD and UCD patients typically present with either compressive symptoms related to local pressure from the mass, like cough, chest pain, and dyspnea or are completely asymptomatic, with a painless, solitary mass often found incidentally (5). Diagnostic workup makes use of CT, which generally shows well-circumscribed mass of soft tissue attenuation with homogenous enhancement in smaller masses and more heterogeneous in appearance in larger lesions (6). On MRI, nodes affected by CD usually demonstrate intermediate to high signal compared to muscle on T1-weighted images and slightly hyperintense on T2-weighted images. Radiological differential diagnoses vary according to location; other vascular tumors like paragangliomas, pheochromocytomas, sarcomas as well as lymphoma must be considered because of their intense enhancement (6). Following these radiologic studies, ultrasound-guided FNA or excisional biopsy may be attempted to achieve a cytologic or tissue diagnosis. Histologically, CD is characterized by nodal expansion that leaves the overall structure of the lymph node largely intact. It may be classified as either the hyaline-vascular or plasma cell variant, with occasional cases demonstrating mixed features (7). The hyaline-vascular variant represents the most common type of CD, showing an increased numbers of lymphoid follicles with features of "regression", a term referring to a predominance of dendritic cells within germinal centers that are hyalinized with a relative paucity of lymphocytes (8). Mantle zones are frequently organized in concentric rings around the germinal center, showing the so-called 'onion-skin' arrangement, a histological finding which is strongly suggestive of CD. Surgical resection provides radical treatment for the majority of patients with UCD (9-10). Radiotherapy is an important alternative for disease that cannot be completely excised. Nevertheless, altough CD is considered a benign disorder, this condition has been associated with an increased risk of developing certain malignancies, as large B-cell lymphomas and follicular dendritic cell sarcomas (3), therefore follow-up is essential.

Conclusion

CD is a rare lymphoproliferative disease that can mimic many malignant conditions. It should be considered in the differential diagnosis when a single persistently nodal mass or multistation lymphadenopathy is associated with intense enhancement. Excisional biopsy is essential to establish a definitive diagnosis to differentiate it from lymphoproliferative disorders and other hypervascular masses.

Conflict of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article

Consent Form for Case Reports: Written informed consent was obtained from the patient concerned.

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