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## Case report

# Unicentric Castleman's disease in the posterior cervical space mimicking a schwannoma



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## ARTICLE INFO

### Keywords:

Castleman's disease  
 Schwannoma  
 Posterior cervical space  
 Hypervascular lesion

## ABSTRACT

**Introduction:** Castleman's disease (CD) is a benign and rare lymphoid tissue disease of undetermined origin. It affects the neck infrequently; therefore its preoperative diagnosis is difficult and can be confused with other hypervascular lesions.

**Case report:** We present a case of unicentric hyaline-vascular CD in the posterior cervical space (PCS) evaluated by MRI and initially presumed to be a cervical schwannoma.

**Discussion:** As shown with our case, unicentric CD should be included in the differential diagnosis for masses in the PCS, especially when a hypervascular lesion like a schwannoma is suspected. The finding of central low intensity areas in a fissured and radial pattern in T2 sequences can help to consider CD when dealing with PCS masses as shown by previous cases and our reported experience.

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## 1. Introduction

Castleman's disease (CD) is a benign and rare lymphoid tissue disease of undetermined origin. It affects the neck infrequently; therefore its preoperative diagnosis is difficult and can be confused with other high vascular lesions like schwannoma. We present a case of unicentric hyaline-vascular CD in the posterior cervical space (PCS) evaluated by MRI and initially presumed to be a cervical schwannoma.

## 2. Case description

A 30-year-old male presented at the head and neck surgery clinic for a slow growing mass located in the left posterior triangle. At physical examination, a single non-pulsatile mass measuring 6 cm in its largest diameter was located in the left posterior cervical triangle below the sternocleidomastoid muscle. It was hard, but was not fixed to deep planes. The palpation elicited no pain and inflammatory changes were absent. With the suspicion of a nerve sheath tumor, a MRI scan was ordered to define extension and primary origin of the lesion. The MRI examination showed a mass in the left

PCS, with an ovoid shape, smooth and well-defined borders and no invasion of adjacent structures, slightly hyperintense to muscle on T1 (Fig. 1A) and hyperintense with scarce central low intensity areas in a radial pattern on T2 (Fig. 1B). After paramagnetic contrast administration, homogeneous enhancement was noted suggesting a hypervascular lesion, possibly a XI cranial nerve or brachial plexus schwannoma. Additionally, two smaller hypervascular nodules inferiorly to the mass were observed (Fig. 2).

At surgery, a neck level IV well-defined non-invasive hypervascular mass was completely resected without complications. The patient was discharged after an uneventful postoperative follow-up. Histologic evaluation of the cervical tumor was consistent with unicentric CD (hyaline vascular variant). Histologic evaluation of the other small nodules observed on MRI was consistent with normal lymph nodes.

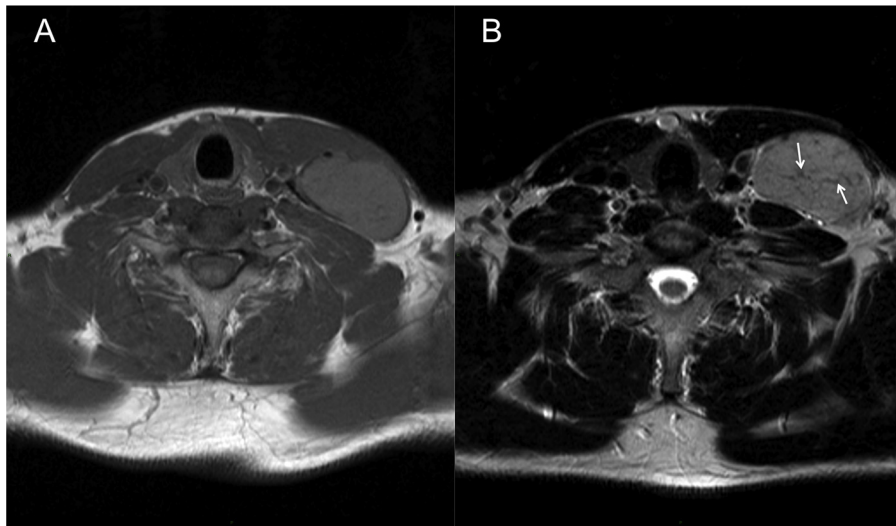
## 3. Discussion

CD is an uncommon condition of unknown etiology affecting lymphoid tissues, without gender preference. Of all cases, 15% occur in the neck. This lesion can present at any age, but unicentric CD is most common in children, adolescents and young adults [1].

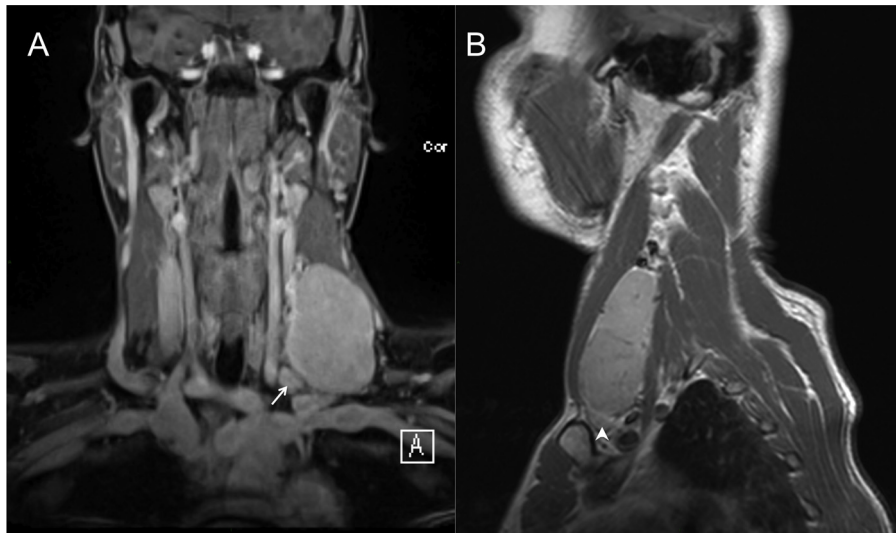
The most common type of CD is the unicentric hyaline-vascular that represents 90% of the cases of CD. It typically manifests as an asymptomatic mass lesion with a benign course and good response

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**Fig. 1.** A. Transverse T1-weighted image shows an ovoid mass in the left PCS, slightly hyperintense with respect to the muscle, homogeneous and with well-defined borders. B. Axial T2-weighted image shows high signal within the mass and central low intensity areas in a fissured and radial pattern (arrows). There is no invasion of adjacent structures.



**Fig. 2.** Coronal T1 VIBE (A) and sagittal T1-weighted (B) gadolinium-enhanced images showing strong and homogeneous enhancement of the mass in the left PCS, deep to the sternocleidomastoid muscle. Note two additional small hypervascular nodules inferomedially (arrow) and inferiorly (arrowhead) to the mass.

to surgical resection. Most lesions in the neck are of this type, as was the lesion in our case [2].

The MRI findings of unicentric hyaline-vascular CD of the neck are similar to CD in other locations. It usually appears as a solitary enlarged lymph node or localized nodal masses, isointense or slightly hyperintense to the muscle on T1-weighted images and hyperintense on T2-weighted images. Intralesional flow voids may be seen on T1- and T2-weighted images reflecting the vascularity of the lesion. Post-contrast T1-weighted images show moderate to marked contrast enhancement [3]. Sometimes, low intensity areas in a fissured and radial pattern may be seen on T2 sequences due to histiocytosis and fibrosis in the central sinusoids [4].

On imaging, the PCS is defined as the area in the posterolateral portion of the neck from the skull base to the clavicles, deep to the sternocleidomastoid and trapezius muscles, but superficial to the perivertebral space. Its main contents are fat, lymph nodes and nerves such as the XI cranial nerve and the components of the brachial plexus, which are the usual origin of masses [5]. Of all PCS

lesions, we think schwannoma merits a special attention because its MRI findings are very similar to CD. It is originated either from XI cranial nerve or roots of the brachial plexus. It tends to be a well-delineated, solitary and fusiform mass, isointense to the muscle on T1-weighted images and hyperintense on T2-weighted images [6]. As in CD, the schwannoma manifests as a hypervascular mass showing homogenous enhancement after contrast administration, but sometimes a cystic area can be identified [7].

#### 4. Conclusion

As shown with our case, unicentric CD should be included in the differential diagnosis for masses in the PCS, especially when a hypervascular lesion like a schwannoma is suspected. Although the differentiation by imaging is difficult, the finding of central low intensity areas in a fissured and radial pattern in T2 sequences can help to consider CD when dealing with PCS masses as shown by previous cases and our reported experience.

### Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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