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Case Report

Video-Assisted Thoracoscopic Safe Minimal Invasive Surgery to Treat Posterior Mediastinal Castleman's Tumor

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Abstract

Castleman disease (CD), referred to as giant lymph node hyperplasia, is a rare lymphoproliferative disorder initially documented by Dr. Benjamin Castleman in 1954. Diagnosis poses a challenge due to its variable clinical presentations and the absence of distinct radiological characteristics. CD can present as either localized (unicentric) or systemic (multicentric), exhibiting diverse histopathological subtypes.

Here we report a new case of unicentric Castleman's disease in the posterior mediastinum presenting with dyspnea and shortness of breath.

Keywords: *Castleman Disease; Castleman Syndrome; Unicentric Castleman's Disease; Multicentric Castleman's Disease; Lymphoproliferative Disorder*

Abbreviation

VATS: Video-Assisted Thoracoscopic Surgery

Introduction

Castleman's disease (CD), alternatively known as giant lymph node hyperplasia or follicular lymphoid hyperplasia, represents a rare and often poorly understood lymphoproliferative disorder. It can manifest wherever lymphoid tissue is present, thus sharing common histological features with lymph nodes [1].

Initially documented by Dr. Benjamin Castleman, Director of the Pathology Department at Massachusetts General Hospital, in a single case in 1954 [2], CD comprises three histological subtypes: hyaline vascular, plasma cellular, and a mixed type, each exhibiting distinct clinical characteristics and survival outcomes [3].

The disease presents two primary clinical manifestations: unicentric Castleman's disease (UCD), characterized by localized signs and symptoms such as isolated lymphadenopathy, and multicentric Castleman's disease (MCD), presenting as a systemic progressive condition with lymphadenopathy affecting multiple sites [4,5].

While most cases of UCD are asymptomatic and incidentally diagnosed through imaging, a minority of patients may experience symptoms related to mass effect [6].

Though the etiology remains unclear, potential factors contributing to CD include viral infections, immune deficiencies, abnormal immune regulation, and dysregulated cytokine secretion. Lymphadenopathy predominantly affects mediastinal sites, accounting for approximately 60% of cases, followed by involvement of retroperitoneal, cervical, and axillary regions [7].

We report a new case of Castleman's disease who presented as a posterior mediastinal mass.

Case Presentation

A 38-year-old female presented with a three-month history of cough, dyspnea, and exertional shortness of breath. Her medical history and physical examination were unremarkable. Routine investigations yielded results within normal limits. Contrast- enhanced CT imaging (Figure 1 and 2) revealed a well-defined lobulated soft tissue density mass with central calcification in the middle mediastinum and subcarinal region.

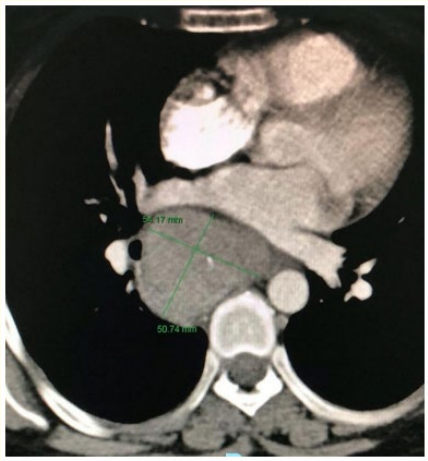

Figure 1: *CT scan with contrast showing soft tissue mass. (Measurements in green).*

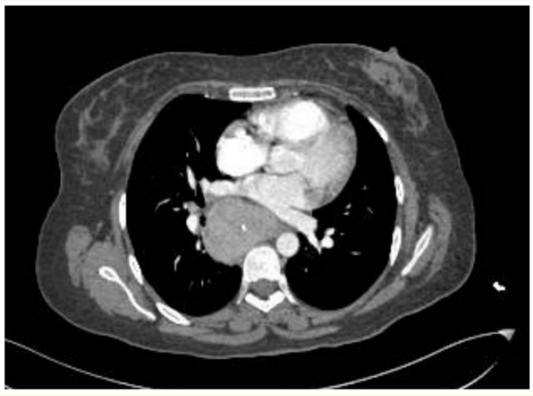

Figure 2: *CT scan with contrast showing soft tissue mass.*

The mass exhibited moderate homogeneous enhancement without signs of adjacent structure invasion. A true-cut biopsy was performed under CT-guidance (Figure 3), but histopathological diagnosis was inconclusive due to insufficient material, prompting a decision to proceed with surgery.

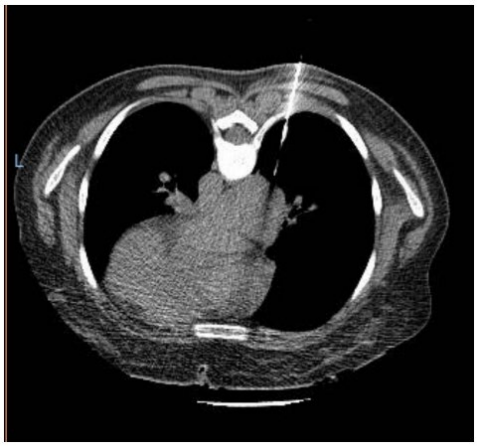

Figure 3: *Patient undergoing true cut biopsy under CT guidance.*

The patient underwent a right posterolateral thoracotomy, revealing an encapsulated mass within the costovertebral sulcus covered by parietal pleura. The mass, located in the paravertebral area of the middle and posterior mediastinum, demonstrated hypervascularity with blood supply derived from intercostal vessels (Figure 4). Complete resection of the tumor was achieved, and the post-operative course was uneventful, with the patient discharged in good general condition.

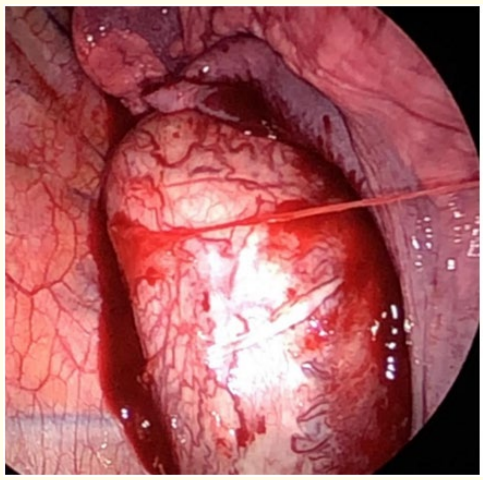

Figure 4: *Screen view of Hyper-vascular mass through VATS.*

Gross and histopathological findings: Histopathological examination of the resected specimen revealed an enlarged lymph node partially effaced by a thick fibrotic capsule. The inter-follicular areas (Figure 5) demonstrated increased cellularity with prominent high endothelial venules. Onion-skinning of the mantle zone was observed (Figure 6). Follicles exhibited variable size and morphology, with regression, central hyalinization, and focal "twinning" (Figure 7).

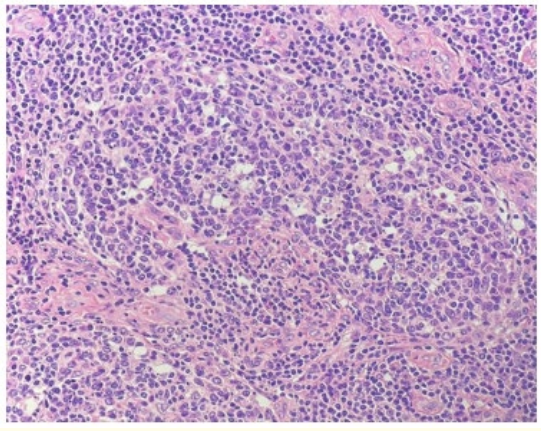

Figure 5: *Interfollicular vascular proliferation.*

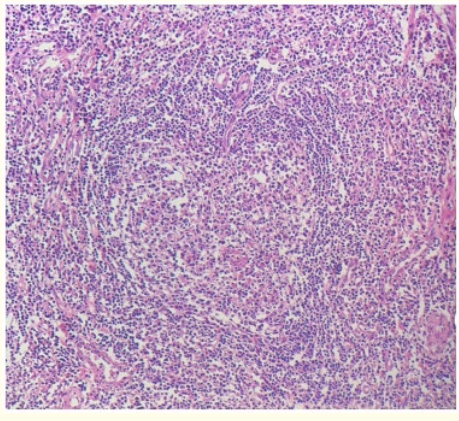

Figure 6: *The follicle is surrounded by thick mantle zone.*

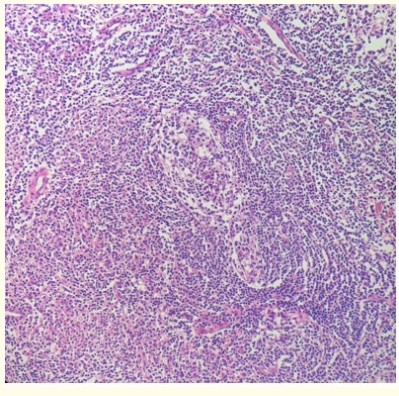

Figure 7: *Twinning of germinal centers and sclerosed vessels.*

Immunohistochemical staining highlighted CD10 and BCL6 within the follicles, underpinned by FDC meshworks positive for CD21 and CD23. CD3 and CD5 highlighted interfollicular T-cells. Negativity was observed for HMB45, CD45, VIMENTIN, EMA, CD68, S100, CD1a, MPO, CD34, cyclin D1, and HHV8.

Discussion

Castleman disease (CD) is a complex condition whose exact cause remains incompletely understood. However, it is known that excessive production of cytokines plays a key role in its development. In unicentric Castleman disease (UCD), interleukin-6 (IL-6) is a crucial factor, while in multicentric Castleman disease (MCD), both IL-6 and human herpesvirus-8 (HHV-8) are clearly involved [8].

CD can affect individuals of any age, though recent studies suggest that UCD tends to occur more often in younger patients, with a slight predominance in women, unlike idiopathic MCD (iMCD) [9].

Castleman disease presents in two main forms: Unicentric Castleman disease (UCD): This form is characterized by localized symptoms, such as isolated lymphadenopathy.

Most cases are asymptomatic and discovered incidentally through imaging. However, some patients may experience symptoms caused by the pressure exerted by enlarged lymph nodes. Multicentric Castleman disease (MCD): This form is a systemic, progressive disease with lymphadenopathy affecting multiple areas [4].

Unlike UCD, MCD can be life-threatening and presents with a more complex range of symptoms. UCD is typically benign and treatable, while MCD can be severe and has four subtypes: idiopathic MCD, HHV-8-positive MCD, TAFRO syndrome, and rarely, MCD associated with POEMS syndrome [9].

Histopathologically, Castleman disease (CD) is classified into three distinct types based on unique lymphoid architectural changes observed in all nodal compartments: hyaline vascular (or hypervascular), plasmacytic, and mixed variants. The hyaline vascular variant is the most common form found in unicentric Castleman disease (UCD), while the plasmacytic variant is more frequently associated with multicentric Castleman disease (MCD). Some cases of MCD also exhibit plasmablastic features [8].

A definitive diagnosis of CD requires histopathological examination of an excised lymph node, as a biopsy is usually not sufficient. Along with histopathology, imaging techniques such as computed tomography (CT) scans of the neck, chest, abdomen, and pelvis, or PET-CT scans, as well as laboratory tests, are necessary to differentiate between UCD and MCD [9].

In radiological evaluation, chest radiographs (CXR) are often the starting point. Typical thoracic CD appears as a rounded solitary mediastinal or hilar mass in asymptomatic patients. On a chest X-ray, mediastinal CD may resemble conditions such as thymoma, lymphoma, or neurogenic tumors, while in some cases, it can mimic bronchial adenomas. Further evaluation with CT imaging reveals that thoracic UCD can be categorized into three morphologic patterns: A solitary, noninvasive mass (seen in 50% of cases), A dominant mass with involvement of adjacent structures (40% of cases), or Matted lymphadenopathy confined to a single mediastinal compartment (10% of cases).

These imaging findings, combined with clinical and laboratory data, help in accurately diagnosing and distinguishing between the types of Castleman disease [10].

Surgical resection is the gold standard for treating unicentric Castleman disease (UCD) and offers a definitive cure for most patients [8]. Minimally invasive thoracoscopic resection is also considered a safe and feasible option, particularly for managing cases of posterior mediastinal Castleman disease [11]. In contrast, treatment for multicentric Castleman disease (MCD) is more complex and often requires

a multifaceted approach. Various therapeutic strategies have been employed, including conventional cytotoxic chemotherapy (either single-agent or combination therapies), antiviral treatments, glucocorticoids, thalidomide, interferon-alpha, and molecular targeted therapies. Thus, the choice of treatment depends on the disease subtype, severity, and individual patient factors [8].

Conclusion

Castleman's disease (CD) is a rare lymphoproliferative disorder with two primary forms: unicentric (UCD), which is localized and often curable through surgical resection, and multicentric (MCD), a systemic condition requiring complex treatments. Video- assisted thoracoscopic surgery (VATS) is a reliable, minimally invasive approach for UCD, offering safer outcomes with reduced recovery times. While UCD has a favorable prognosis, MCD is more challenging and necessitates a multifaceted treatment strategy based on disease severity and subtype.

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