

Scrotal Hernia or Castleman Disease? A Diagnostic Pitfall in an Unusual Presentation of a Rare Entity

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ABSTRACT

Castleman disease (CD) is a rare, non-neoplastic lymphoproliferative disorder with varied clinical presentations and uncertain etiology. It is classified into unicentric (UCD) and multicentric (MCD) forms, and histologically into hyaline vascular, plasma cell, or mixed subtypes. While CD typically affects the mediastinum or neck, scrotal involvement is exceedingly rare and can mimic more common conditions. We report a case of a 65-year-old man with a painless, enlarging scrotal mass initially diagnosed as a reducible hernia. Imaging and histopathology revealed a large lymphoid mass, diagnosed as UCD with mixed hyaline vascular and plasma cell subtypes. This case highlights the importance of including CD in the differential diagnosis of scrotal masses and underscores the critical role of histopathological examination in diagnosis and treatment planning. The patient had a favorable outcome after surgical excision, reinforcing that early recognition and proper management of UCD typically result in an excellent prognosis.

Keywords

Unicentric Castleman disease, scrotal mass, histopathology, surgical excision, mixed subtype

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Received: 22nd April 2025; Accepted: 14th
February 2026

Doi:
<https://doi.org/10.31436/10.31436/imjm.v25i04/2943>

INTRODUCTION

Castleman disease (CD) is a non-neoplastic lymphoproliferative disorder first described by Benjamin Castleman in 1954. Although considered benign, CD poses a diagnostic challenge due to its highly variable presentation and unclear etiology. CD is clinically classified into two main forms: Unicentric Castleman Disease (UCD), which involves a single lymph node region, and Multicentric Castleman Disease (MCD), which affects multiple lymph node regions with systemic manifestations. Histopathologically, CD is categorized into three subtypes: hyaline-vascular, plasma cell, and mixed type, with the hyaline-vascular variant being the most common.^{1,2}

CD most frequently arises in the mediastinum (31%), the neck (42%), the abdomen (18%), and the retroperitoneum (5%). Involvement of the scrotal region

is extremely rare and scarcely reported in the literature, making it a diagnostic challenge that can be easily mistaken for a more common condition, such as scrotal hernia.³ CD may present as a reducible mass, clinically resembling a hernia, thus necessitating a thorough differential diagnosis in such presentations.

While UCD predominantly occurs in the mediastinum, cervical, abdominal, or retroperitoneal nodes, scrotal involvement is exceedingly rare and scarcely documented. This rarity contributes to frequent misdiagnosis, particularly as inguinal or scrotal hernia-conditions far more common in clinical settings. Hernias, particularly of the indirect type, account for a lifetime risk of 27% in men.⁴ The overlapping clinical features, such as a reducible mass in the groin, further complicate diagnostic accuracy. Unlike typical hernias,

CD does not transilluminate and may not reduce fully, though these distinctions may be subtle.

The case presented here is a rare instance of UCD with a mixed histological subtype (hyaline-vascular and plasma cell), manifesting as a scrotal mass in a patient with no systemic symptoms. The unusual anatomical location and histological variant underscore the novelty of this case, highlighting the importance of considering CD in atypical scrotal masses. Histopathological examination remains the gold standard for diagnosis and therapeutic planning, with complete surgical excision typically curative in UCD cases.^{5,6}

CASE REPORT

A 65-year-old male with a history of previously treated pulmonary tuberculosis presented with a progressively enlarging scrotal mass, which he had first noticed approximately one year prior. The swelling was bilateral, painless, and gradually increased in size over time. Notably, he did not report any systemic symptoms such as fever, weight loss, or night sweats.

Initially, the patient consulted a general practitioner who suspected a bilateral, reducible scrotal hernia based on the partially reducible nature of the mass on physical exam. There were no gastrointestinal or genitourinary complaints. The patient had no prior significant surgical history, and baseline laboratory investigations, including complete blood count, renal and liver function tests, as well as tumor markers (AFP, β -HCG, LDH), were within normal limits.

Physical Examination, on inspection, bilateral scrotal enlargement was observed, with the left side more prominently affected. There was no palpable swelling above the pubic tubercle, a non-tender mass without transillumination, and the upper border of the mass could be clearly identified. This clinical finding was what initially led to the suspicion that the mass might not be a simple indirect hernia. Partial reducibility on examination reinforced the initial suspicion of an inguinal hernia.

Scrotal ultrasonography showed a heterogeneous mass with internal vascularity, suggesting a solid lesion rather than herniated bowel content. The ultrasound was performed to differentiate the atypical mass from a standard hernia by ruling out solid tumors or other scrotal pathologies. Despite this, the clinical team maintained a working diagnosis of incarcerated hernia and scheduled the patient for elective surgical exploration and herniotomy.

Intraoperatively, the surgical team encountered unexpected large lymphadenopathy within the left inguinal canal. No bowel herniation was found. The lymph nodes appeared firm and white on the cut surface. Macroscopic tissue mass is three pieces of enlarged lymph node, dimensions 8x7,7x8,5 cm; 3,5x3x2 cm; 4,5x2,5x1,5 cm, firm and white cut surface (figure 1). A standard herniotomy was performed as planned. Notably, the mass adhered to the spermatic cord, requiring meticulous dissection to avoid damage to adjacent structures. No frozen section was available, so the tissue was submitted for formal histopathological evaluation.

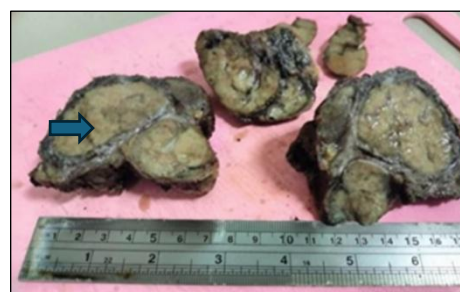


Figure 1: Macroscopic Mass in CD

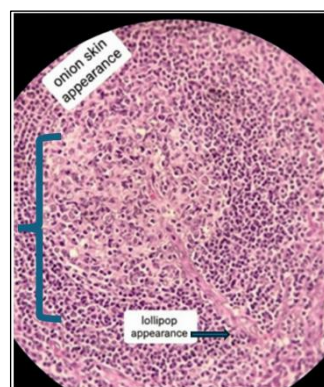


Figure 2: Onion skin appearance and lollipop appearance in CD

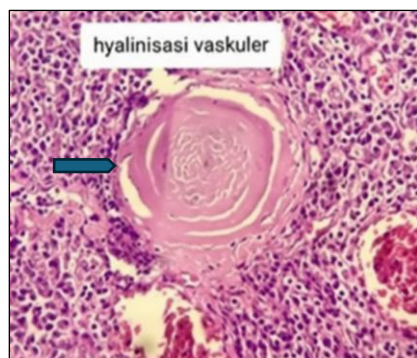


Figure 3. Hyalin Vascular in CD

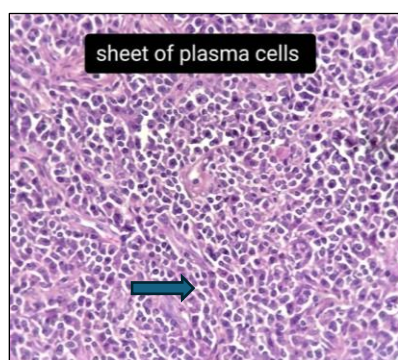


Figure 4. Plasma cell in CD

Histopathology and Final Diagnosis, microscopic examination revealed features diagnostic of Castleman disease, mixed hyaline vascular and plasma cell type. Histological hallmarks included: thickened fibrous capsule and marked follicular hyperplasia, hyalinized and sclerotic blood vessels traversing involuted germinal centers, producing a characteristic “lollipop” appearance, Concentric layering of lymphocytes in the mantle zones (“onion-skinning”) (figure 2) Interfollicular areas rich in mature plasma cells (figure 3), with prominent vascular proliferation and perivascular hyalinization (figure 4).

The final diagnosis was Unicentric Castleman Disease (UCD) of mixed histopathological subtype, involving the scrotal region an exceedingly rare anatomical location for this condition.

DISCUSSION

Scrotal hernia is among the most common conditions encountered in surgical practice, with an estimated lifetime risk of 27% in men and approximately 3% in

women. Indirect inguinal hernias occur twice as frequently as direct types, and right-sided hernias are more prevalent than left-sided ones. A significant male predominance is well-documented, with a male-to-female ratio of 7:1.^{7,8} Given its frequency and clinical familiarity, a scrotal hernia is often the initial diagnosis when encountering reducible scrotal masses.

However, Castleman disease (CD), a rare and poorly understood lymphoproliferative disorder, may masquerade as more common scrotal pathologies, including hernias. CD is clinically classified into unicentric (UCD), often presenting as a localized mass (commonly hyaline-vascular type), and multicentric (MCD), which is associated with systemic symptoms and typically plasma-cell histology.^{2,9} Most UCD cases involve the mediastinum (31%), neck (42%), abdomen (18%), or retroperitoneum (5%). While over 500 cases of CD have been reported globally, documented scrotal involvement remains extraordinarily rare, with fewer than two cases described in the English literature to date.³

Unicentric Castleman disease typically affects mediastinal or thoracic lymph nodes, with patients often being asymptomatic and cured following complete surgical excision. In contrast, MCD presents with generalized lymphadenopathy, constitutional symptoms (fever, night sweats, weight loss), and elevated inflammatory markers, and requires systemic therapy rather than surgery.¹⁰

The etiopathogenesis of CD remains elusive, though dysregulated cytokine production particularly interleukin-6 (IL-6) has been implicated. HHV-8 infection is commonly associated with MCD, especially in immunocompromised patients.⁹⁻¹⁰

In this case, a scrotal mass presumed to be a hernia revealed itself intraoperatively as enlarged lymph nodes, which were histologically confirmed as UCD of mixed hyaline-vascular and plasma-cell subtype. Pain and activity-related discomfort, uncommon in localized CD

added to the diagnostic confusion. These atypical features underscore the need for high clinical suspicion and comprehensive evaluation of scrotal masses, particularly when standard imaging or physical findings are incongruent. The scrotum remains an exceptionally unusual site, often leading to misdiagnosis, as symptoms and physical findings may resemble those of more common conditions such as hernias, hydrocele, or testicular tumors.

Complete surgical excision remains the definitive treatment for UCD, offering an excellent prognosis with minimal recurrence. In contrast, MCD typically requires systemic treatment such as corticosteroids, antiviral agents (e.g., ganciclovir), monoclonal antibodies (rituximab, tocilizumab), or immunomodulatory therapy due to its more aggressive clinical course.^{2,9,10}

CONCLUSION

This case highlights a rare presentation of UCD in the scrotal region, initially suspected as a bilateral reducible scrotal hernia. When a scrotal mass presents with atypical clinical signs, early imaging evaluation is crucial to avoid diagnostic pitfalls. Intraoperative findings revealed isolated lymphadenopathy in the inguinal canal, confirmed by histopathology as a mixed hyaline-vascular and plasma cell variant of CD. Notably, pain and activity-dependent discomfort unusual for UCD were present, further complicating the clinical picture. These findings underscore the importance of including rare etiologies such as Castleman disease in the differential diagnosis of atypical scrotal masses.

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