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**Case Report**

# Unicentric Castleman Disease of the Neck Mimicking Lymphoma: A Diagnostic Challenge Highlighting the Role of Histopathology and Immunohistochemistry

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## ABSTRACT

Castleman disease is a rare, benign lymphoproliferative disorder that often poses a diagnostic dilemma due to its clinical and radiological resemblance to lymphoma. We report a case of unicentric Castleman disease of the cervical lymph node in a young patient, emphasizing the importance of histopathological examination and immunohistochemistry in establishing a definitive diagnosis. The patient presented with a long-standing, painless cervical lymphadenopathy without constitutional symptoms. Radiological evaluation revealed an isolated enlarged lymph node, raising suspicion of a neoplastic lymphoid disorder. Fine needle aspiration cytology showed a polymorphous lymphoid population but was inconclusive.

Excision biopsy demonstrated partial effacement of nodal architecture with numerous small regressed germinal centers surrounded by concentric layers of mantle zone lymphocytes, imparting an "onion-skin" appearance. Prominent hyalinized blood vessels penetrating the follicles were noted, forming characteristic "lollipop" lesions. Interfollicular areas showed increased vascularity without sheets of plasma cells. On immunohistochemistry, lymphoid follicles were highlighted by CD20, interfollicular T cells were CD3 positive, BCL2 showed positivity in mantle zone lymphocytes, CD5 showed focal interfollicular positivity, and Cyclin D1 was negative, effectively excluding mantle cell lymphoma.

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A final diagnosis of unicentric Castleman disease, hyaline vascular type, was rendered. The patient underwent complete surgical excision with no recurrence on follow-up. This case underscores the importance of considering Castleman disease in the differential diagnosis of isolated cervical lymphadenopathy and highlights the pivotal role of histomorphology and immunohistochemistry in avoiding misdiagnosis and overtreatment.

**KEYWORDS**

• Castleman disease • Unicentric • Hyaline vascular type • Cervical lymphadenopathy • Immunohistochemistry • Lymphoproliferative disorder

## INTRODUCTION

Castleman disease (CD) is a rare, benign lymphoproliferative disorder characterized by non-clonal lymph node hyperplasia, First described by Castleman in 1956.[1] It is broadly classified into unicentric Castleman disease (UCD) and multicentric Castleman disease (MCD), with the unicentric form most commonly exhibiting the hyaline vascular subtype.[2] UCD typically presents as an isolated lymph node enlargement and is often asymptomatic, whereas MCD is associated with systemic inflammatory manifestations and poorer prognosis.[3]

Pediatric Castleman disease is particularly uncommon, and cervical involvement in children is even rarer, resulting in frequent misdiagnosis as lymphoma, tuberculosis, or reactive lymphadenopathy.[4] Radiological findings are nonspecific, and fine needle aspiration cytology may be inconclusive, making histopathological examination with immunohistochemistry essential for accurate diagnosis.[5]

We report the case of a **10-year-old boy** presenting with a **one-month history** of painless left cervical lymphadenopathy. Laboratory investigations revealed **elevated inflammatory markers (ESR and CRP),** while **HIV serology was non-reactive.** Radiological evaluation demonstrated an isolated enlarged cervical lymph node, raising suspicion for a lymphoid neoplasm. Fine-needle aspiration cytology was inconclusive, prompting excision biopsy. Histopathological examination revealed regressed germinal centres with concentric mantle zone lymphocytes ("onion-skin" appearance), hyalinized penetrating vessels ("lollipop" lesions), and increased interfollicular vascularity. Immunohistochemistry demonstrated CD20-positive follicles, CD3-positive interfollicular T lymphocytes, BCL2 positivity in mantle zone lymphocytes, focal CD5 positivity, and negative Cyclin D1 expression, thereby excluding mantle cell lymphoma. A diagnosis of **unicentric Castleman disease, hyaline vascular type**, was established. The patient underwent complete surgical excision and remained disease-free during **6 months of follow-up.**

Surgical excision alone is curative in unicentric disease, and timely diagnosis significantly alters patient management and prognosis.[6] Reporting such cases contributes to improved awareness among clinicians and pathologists and expands the limited pediatric literature on this rare entity.[7]

### Case Presentation

A **10-year-old boy** presented with a **one-month history** of a painless swelling on the left side of the neck. There was no history of fever, weight loss, night sweats, or other constitutional symptoms. Physical examination revealed **a firm, mobile, well-circumscribed, non-tender left cervical lymph node measuring approximately 3 × 3 cm.** No additional lymphadenopathy or hepatosplenomegaly was detected.

Laboratory investigations demonstrated **elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP)**, suggesting an inflammatory process. Routine hematological and biochemical investigations were otherwise within normal limits. **HIV serology was non-reactive, while human herpesvirus-8 (HHV-8) testing was not performed.**

Ultrasonography demonstrated a well-defined enlarged cervical lymph node with increased vascularity. Contrast-enhanced computed tomography revealed an isolated

intensely enhancing left level II cervical lymph node, raising suspicion for lymphoma. Fine-needle aspiration cytology demonstrated a polymorphous lymphoid population but was inconclusive. Owing to persistent diagnostic uncertainty, complete excision of the lymph node was performed for histopathological evaluation.

### Histopathological Findings

Microscopic examination showed partial effacement of lymph node architecture. Numerous small follicles with regressed germinal centers were present. These follicles were surrounded by concentric layers of mantle zone lymphocytes, producing the classic "onion-skin" pattern. Hyalinized blood vessels penetrating the germinal centers resulted in the characteristic "lollipop" appearance. The interfollicular regions were moderately expanded with prominent vascular proliferation but lacked sheets of plasma cells or atypical lymphoid infltrates Immunohistochemical analysis demonstrated strong CD20 expression within the lymphoid follicles, while CD3 highlighted interfollicular T lymphocytes. BCL2 expression was confined to mantle zone lymphocytes with relative sparing of the germinal centres. CD5 showed focal positivity in interfollicular lymphocytes, and Cyclin D1 expression was absent, effectively excluding mantle cell lymphoma. These findings, together with the characteristic histomorphological features, confirmed the diagnosis of u**nicentric Castleman disease, hyaline vascular type.**

### Treatment and Follow-Up

The patient underwent complete surgical excision of the affected cervical lymph node. The postoperative course was uneventful. During **6 months of clinical follow-up**, the patient remained asymptomatic, with no clinical or radiological evidence of disease recurrence.

## DISCUSSION

Pediatric Castleman disease is sparsely reported in the literature, with most cases presenting as unicentric hyaline vascular type involving the mediastinum or cervical lymph nodes. The present case shares several features with previously reported pediatric cases, including absence of systemic symptoms, localized disease, and excellent outcome following surgical excision.

The present patient was a **10-year-old immunocompetent boy** with localized cervical lymphadenopathy of one month's duration. Although inflammatory markers (ESR and CRP) were elevated, there were no constitutional symptoms or laboratory evidence of systemic disease. HIV serology was non-reactive, and HHV-8 testing was not performed because the clinical and histopathological findings were characteristic of unicentric hyaline vascular Castleman disease. These findings are consistent with previously reported pediatric cases, in which complete surgical excision is both diagnostic and curative.

**Table 1:** Recent Pediatric / Unicentric castleman disease cases including present case

| Author (Year) | Age | Site | Type | Key Pathology / IHC | Treatment | Outcome |
|---|---|---|---|---|---|---|
| **Bi *et al.* (2024)**[7] | Child | Cervical | UCD (HV) | Onion-skin follicles, hyalinized vessels; CD20+, CD3+ | Excision | No recurrence |
| **van Rhee *et al.* (2020)**[2] | All ages | Nodal | UCD | Typical HV morphology; IHC to exclude lymphoma | Surgery | Excellent |
| **Wang J *et al.* (2020)**[11] | Pediatric | Various | UCD > MCD | Classical HV features; favorable pathology | Excision | Good |
| **Zhang *et al.* (2015)**[8] | Children | Multisite | Predominantly UCD | HV type common; CD20+, CD3+ | Excision | Disease-free |
| **Present case** | Pediatric | Cervical | UCD (HV) | Onion-skin mantle zones, lollipop lesions; CD20+, CD3+, BCL2+, focal CD5+, Cyclin D1− | Excision | No recurrence |

### Interpretation of the Comparative Data

Recent literature underscores that pediatric Castleman disease is rare, with unicentric involvement being the predominant form and most commonly affecting the mediastinum or cervicallymphnodes. Arecentclinicopathologic

study by Bi *et al.* demonstrated that pediatric unicentric Castleman disease of the neck typically presents without systemic symptoms and shows characteristic histomorphological and immunohistochemical features, allowing definitive diagnosis on excision biopsy.[7] International evidence-based consensus guidelines emphasize that complete surgical excision is curative for unicentric Castleman disease and should be considered the treatment of choice.[2] Pediatric case series further support excellent long-term outcomes following surgery alone, with minimal risk of recurrence.[11] Advances in understanding disease biology have clarified the cytokine-driven pathogenesis of multicentric Castleman disease, aiding distinction from unicentric forms.[3] Updated pathological classifications and diagnostic criteria remain essential to differentiate Castleman disease from malignant lymphomas, thereby preventing unnecessary aggressive therapy.[9] Across reported pediatric cases, unicentric Castleman disease consistently demonstrates:

- Predominance of hyaline vascular subtype
- Localized disease without systemic symptoms
- Excellent prognosis following complete surgical excision

The present case aligns with these observations, reinforcing surgery as definitive therapy and highlighting the importance of recognizing this entity in children presenting with isolated lymphadenopathy.[10,11]

Unicentric Castleman disease, particularly the hyaline vascular type, commonly presents as an isolated lymph node enlargement and is often asymptomatic. The neck is the second most common site after the mediastinum. Radiological features, though suggestive, are not specific and often overlap with malignant conditions.

Histopathology remains the gold standard for diagnosis. The presence of regressed germinal centers, onion-skin mantle zones, and hyalinized penetrating vessels are hallmark features. Immunohistochemistry plays a critical role in excluding lymphomas, especially mantle cell lymphoma and follicular lymphoma, which can closely mimic CD morphologically.

### Why this Case is Important to Publish

This case is important because:

1. Castleman disease is rare and frequently misdiagnosed.

It closely mimics lymphoma clinically and radiologically, leading to potential overtreatment.

1. Accurate histopathological and immunohistochemical diagnosis prevents unnecessary chemotherapy.
2. It reinforces surgical excision as curative therapy in unicentric disease.
3. It contributes to awareness among pathologists and clinicians, particularly in resource-limited settings.

## CONCLUSION

Unicentric Castleman disease should be considered in the differential diagnosis of isolated cervical lymphadenopathy in children. Careful histomorphological evaluation supplemented by appropriate immunohistochemistry remains the cornerstone for establishing an accurate diagnosis and excluding malignant lymphoma. Complete surgical excision provides both definitive diagnosis and curative treatment, resulting in an excellent prognosis with a low risk of recurrence.

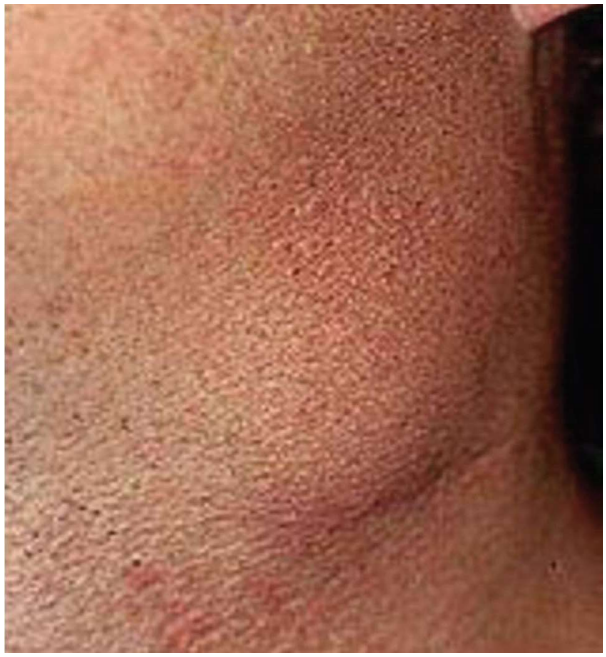

**Figure 1:** Clinical photograph showing a solitary, well-defined swelling over the left side of the neck measuring approximately 3 × 3 cm. The swelling was firm, mobile, well-marginated, and non-tender on palpation

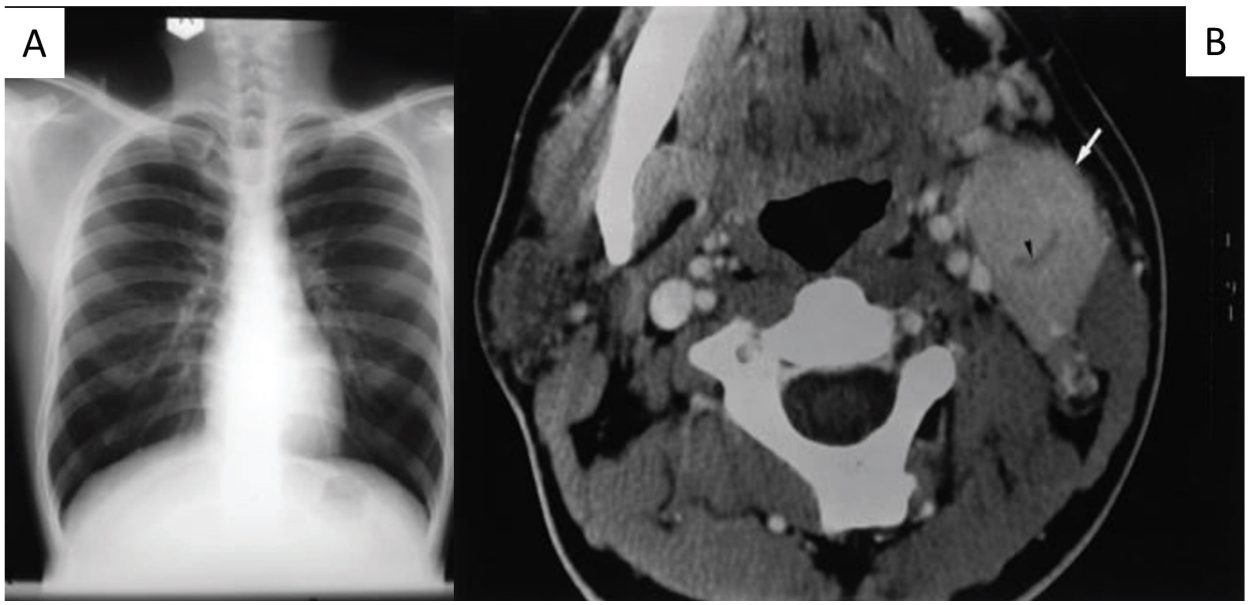


**Figure 2:** (**A)** Chest radiograph (posteroanterior view) showing normal lung fields and mediastinal contours, with no evidence of mediastinal widening, pulmonary lesion, or lymphadenopathy; **(B)** Axial contrast-enhanced computed tomography (CT) scan of the neck demonstrating an enlarged, well-enhancing left level II cervical lymph node (white arrow) with a central crescentic low-attenuation band (arrowhead), a radiological feature suggestive of a hypervascular lymphoid lesion.

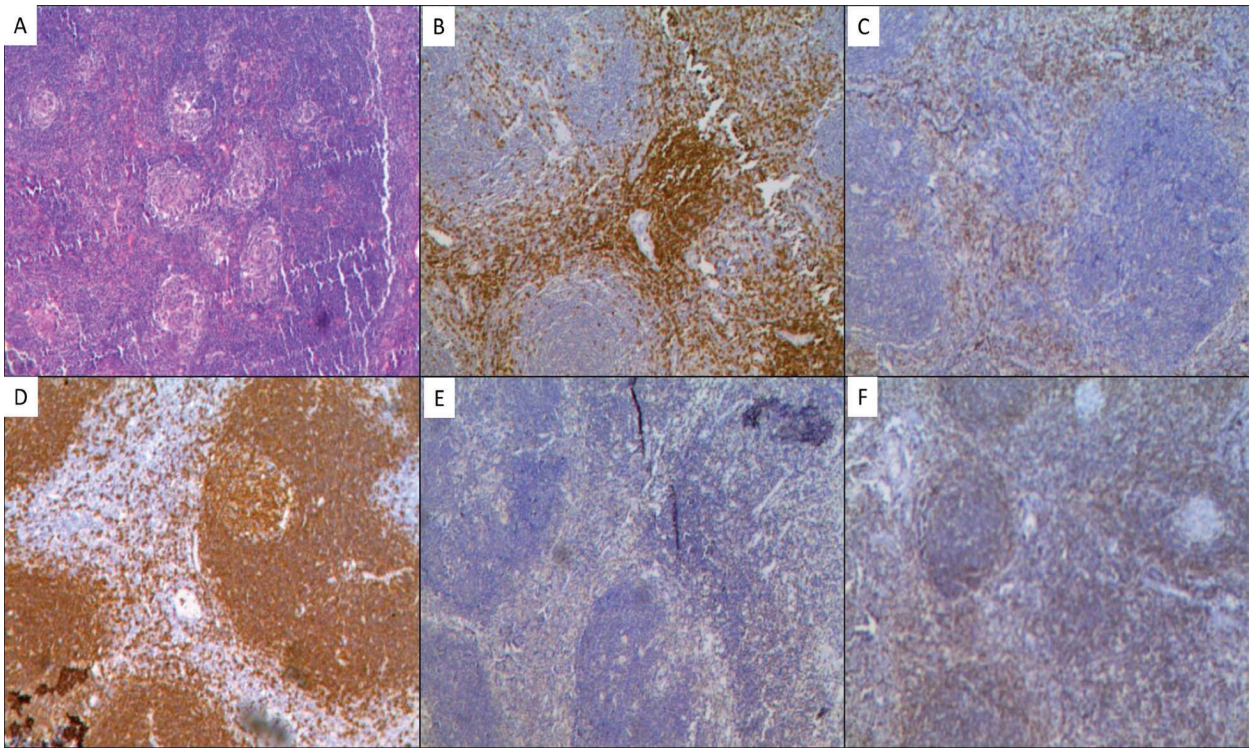


**Figure 3:** **(A)** Hematoxylin and eosin–stained section showing partial effacement of lymph node architecture with multiple regressed germinal centers surrounded by concentric layers of mantle zone lymphocytes, imparting an "onion-skin" appearance, along with prominent hyalinized blood vessels consistent with hyaline vascular type Castleman disease; **(B)** CD3 immunohistochemistry highlighting interfollicular T lymphocytes; **(C)** CD5 immunohistochemistry showing focal positivity in interfollicular lymphocytes; **(D)** CD20 immunohistochemistry demonstrating strong positivity in lymphoid follicles; **(E)** Cyclin D1 immunohistochemistry showing negative staining, excluding mantle cell lymphoma; **(F)** BCL2 immunohistochemistry showing positivity in mantle zone lymphocytes with relative sparing of germinal centers.

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