Case Reports in Clinical Medicine, 2026, 15(4), 133-138
https://www.scirp.org/journal/crcm
ISSN Online: 2325-7083
ISSN Print: 2325-7075

Unicentric Castleman Disease Presenting as an Isolated Submandibular Mass: Diagnostic Challenges for the Otolaryngologist

Nawress Thabet[1*], Jihene Houas[1], Maissa Thabet[2], Malika El Omri[1], Nihed Abdessayed[3], Neirouz Ghannouchi[2]

[1]ENT Department, Farhat Hached University Hospital, Faculty of Medicine of Sousse, Sousse, Tunisia
[2]Internal Medicine Department, Farhat Hached University Hospital, Faculty of Medicine of Sousse, Sousse, Tunisia
[3]Pathology Department, Farhat Hached University Hospital, Faculty of Medicine of Sousse, Sousse, Tunisia
Email: *nawress.thabet@hotmail.com

How to cite this paper: Thabet, N., Houas, J., Thabet, M., El Omri, M., Abdessayed, N. and Ghannouchi, N. (2026) Unicentric Castleman Disease Presenting as an Isolated Submandibular Mass: Diagnostic Challenges for the Otolaryngologist. *Case Reports in Clinical Medicine*, **15**, 133-138.
https://doi.org/10.4236/crcm.2026.154018

Received: March 9, 2026
Accepted: March 28, 2026
Published: March 31, 2026

Abstract

Background: Castleman disease (CD) is a rare lymphoproliferative disorder characterized by abnormal lymph node hyperplasia. Although the mediastinum represents the most frequent site of involvement, cervical localization is uncommon and may present a diagnostic challenge due to nonspecific clinical and radiological findings. **Case Presentation:** We report the case of a 44-year-old woman who presented with a progressively enlarging painless mass in the left submandibular region evolving over two months. Physical examination revealed a firm, mobile lymph node measuring approximately 2 cm without associated systemic symptoms. Laboratory investigations were unremarkable. Cervical computed tomography demonstrated a well-defined lymph node enlargement at level IB. The patient underwent complete surgical excision of the lesion. Histopathological examination revealed regressed germinal centers with hyalinized vessels and concentric mantle zones ("onion-skin" appearance), consistent with the hyaline-vascular subtype of unicentric Castleman disease. Postoperative positron emission tomography showed no additional sites of disease. **Discussion:** Cervical unicentric Castleman disease is rare and often mimics other causes of cervical lymphadenopathy, including lymphoma, metastatic lymph nodes, or benign inflammatory conditions. Because imaging and cytology are frequently inconclusive, definitive diagnosis relies on histopathological evaluation following surgical excision. **Conclusion:** This case highlights the importance of considering Castleman disease in the differential diagnosis of isolated cervical lymphadenopathy. Complete surgical excision remains both the diagnostic and curative treatment for unicentric forms of the disease.

Keywords

Castleman Disease, Cervical Lymphadenopathy, Unicentric Castleman Disease, Neck Mass, Case Report

1. Introduction

Castleman disease (CD) is a rare benign lymphoproliferative disorder first described by Benjamin Castleman in 1954 [1]. It is characterized by lymph node hyperplasia with specific histological features [2]. Although CD may involve any lymph node station, approximately 70% of cases occur in the mediastinum, while cervical and abdominal localizations account for nearly 15% of cases [3].

Clinically, CD is classified into unicentric Castleman disease (UCD), involving a single lymph node region, and multicentric Castleman disease (MCD), characterized by generalized lymphadenopathy and systemic inflammatory manifestations. Advances in disease classification and pathogenesis have clarified the heterogeneity of CD and emphasized the role of inflammatory cytokines and viral factors in its development [3].

We report a case of unicentric Castleman disease located in the cervical region and review recent advances in diagnosis and management.

2. Case Presentation

A 44-year-old woman with no significant medical history presented with a progressively enlarging, painless left submandibular mass evolving over two months. She denied fever, night sweats, weight loss, asthenia, dysphagia, dysphonia, or dyspnea.

Physical examination revealed a firm, mobile, non-tender mass measuring approximately 2 cm in diameter in the left submandibular region. The overlying skin was normal. Examination of the ear, nose, nasopharynx, oropharynx, and larynx showed no abnormalities. No additional lymphadenopathy was detected.

Routine laboratory tests, including inflammatory markers, were within normal limits. Cervical computed tomography demonstrated a well-defined enlarged lymph node at level IB (**Figure 1**).

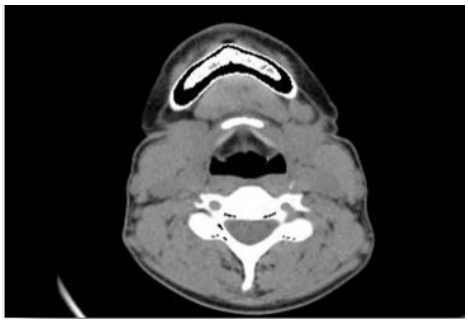

Figure 1. Axial cervical CT scan showing an enlarged lymph node at the left level IB.

Fine-needle aspiration cytology was performed but yielded inconclusive results. Due to persistent diagnostic uncertainty, complete surgical excision was performed under general anesthesia. Histopathological examination demonstrated regressed germinal centers with hyalinized vessels penetrating the follicles, concentric layering of small lymphocytes ("onion-skin" appearance), and prominent vascular proliferation—findings consistent with the hyaline-vascular subtype of unicentric Castleman disease (Figure 2). Immunohistochemical analysis did not support lymphoma or other lymphoproliferative disorders, thereby confirming the diagnosis.

To establish unicentric disease, systemic evaluation including complete blood count, inflammatory markers, and viral screening (including HIV and HHV-8 where applicable) showed no abnormalities. A postoperative PET scan confirmed the absence of additional disease sites (Figure 3).

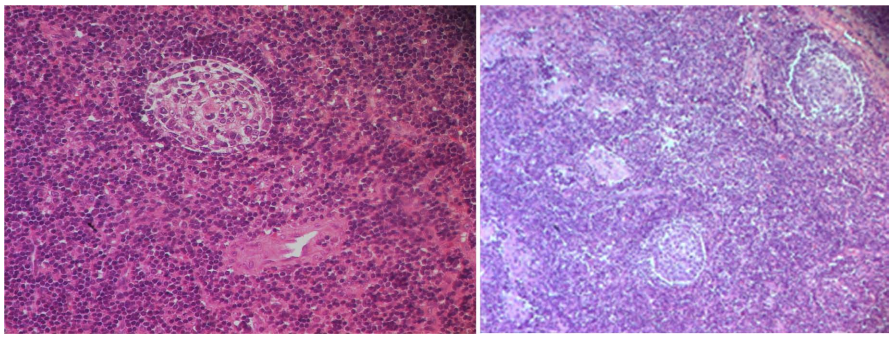

Figure 2. Histopathology of cervical lymph node showing the hyaline-vascular type of Castleman disease with concentric mantle zones ("onion-skin" appearance) and hyalinized vessels.

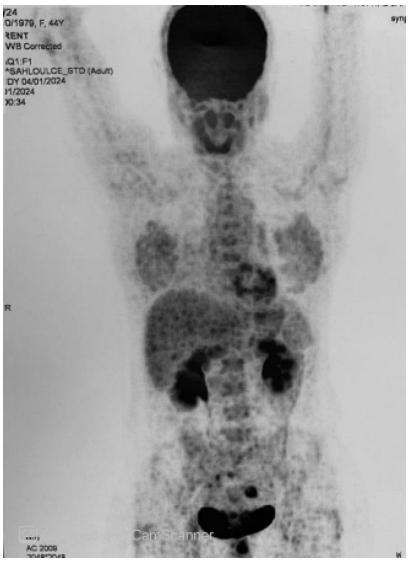

Figure 3. Postoperative PET scan showing no abnormal metabolic activity.

The postoperative course was uneventful. At 12-month follow-up, the patient remained asymptomatic with no evidence of recurrence.

3. Discussion

Castleman disease (CD) is a rare lymphoproliferative disorder characterized by benign lymph node hyperplasia. Although the mediastinum is the most frequently affected site, head and neck involvement represents approximately 10% - 15% of reported cases, with cervical lymph nodes being the most common location in this region. Because of its rarity and nonspecific clinical presentation, cervical Castleman disease may pose a diagnostic challenge for otorhinolaryngologists and is often initially suspected to be another cause of cervical lymphadenopathy [3]-[5].

Two clinical forms of CD are recognized: unicentric Castleman disease (UCD) and multicentric Castleman disease (MCD). UCD involves a single lymph node region and is usually asymptomatic, presenting as a slowly enlarging solitary mass. In contrast, MCD is characterized by generalized lymphadenopathy and systemic inflammatory symptoms such as fever, weight loss, and fatigue [4] [6]. The absence of systemic manifestations and the presence of isolated cervical lymphadenopathy in our patient were consistent with the unicentric form.

In the head and neck region, cervical lymph nodes are the most commonly affected sites. However, due to its rarity and nonspecific presentation, cervical Castleman disease is frequently misdiagnosed preoperatively. The differential diagnosis includes reactive lymphadenopathy, lymphoma, metastatic lymph nodes, salivary gland tumors, and congenital cystic lesions such as branchial cleft cysts. Imaging modalities such as computed tomography or magnetic resonance imaging typically demonstrate a well-circumscribed and hypervascular lymph node, but these findings are not pathognomonic [5] [7].

Fine-needle aspiration cytology is frequently inconclusive and may lead to misdiagnosis, particularly as lymphoma [8] [9]. Therefore, complete surgical excision is usually required for definitive diagnosis.

Histopathological examination remains the gold standard for diagnosis. Three histological variants are described: hyaline-vascular, plasma cell, and mixed types. The hyaline-vascular subtype is the most common form of unicentric disease and accounts for the majority of head and neck cases. [10] It is characterized by regressed germinal centers, concentric mantle zones producing the classic "onion-skin" appearance, and prominent vascular proliferation [2] [11]. These features were observed in the present case.

Recent advances in Castleman disease have highlighted the role of interleukin-6 (IL-6) in disease pathogenesis, particularly in multicentric forms. Targeted therapies such as anti-IL-6 agents have significantly improved outcomes in MCD [12] [13], while unicentric disease continues to be effectively managed with surgical excision alone [14].

In the present case, the absence of systemic symptoms, normal laboratory findings, and negative imaging confirmed the unicentric form. This distinction is critical, as management strategies differ substantially.

Preoperative diagnosis remains challenging. Imaging findings are nonspecific, and fine-needle aspiration often fails to provide a definitive diagnosis. Therefore,

surgical excision is both diagnostic and therapeutic in most cases of suspected UCD.

4. Conclusion

Castleman disease is a rare benign lymphoproliferative disorder that may present as an isolated cervical mass, posing diagnostic challenges. Histopathological examination remains essential for definitive diagnosis. Unicentric Castleman disease is effectively treated by complete surgical excision, which provides excellent prognosis and low recurrence rates. Increased awareness among otolaryngologists is crucial to ensure accurate diagnosis and appropriate management.

Ethics Statement

Written informed consent was obtained from the patient for publication of this case report and accompanying images. According to institutional policy, ethics committee approval was not required for single case reports.

Acknowledgements

The authors thank the medical and surgical teams involved in the patient's care.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Shahidi, H., Myers, J.L. and Kvale, P.A. (1995) Castleman's Disease. *Mayo Clinic Proceedings*, **70**, 969-977. https://doi.org/10.4065/70.10.969

[2] Keller, A.R., Hochholzer, L. and Castleman, B. (1972) Hyaline-Vascular and Plasma-Cell Types of Giant Lymph Node Hyperplasia of the Mediastinum and Other Locations. *Cancer*, **29**, 670-683.

[3] Lin, C. and Chang, Y. (2010) Castleman's Disease in the Head and Neck Region: Meta-Analysis of Reported Cases in Taiwan Region and Literature Review. *Journal of the Formosan Medical Association*, **109**, 913-920. https://doi.org/10.1016/s0929-6646(10)60139-8

[4] Brúgós, B., Simon, Z., Méhes, G., Illés, Á. and Pfliegler, G. (2024) Diagnostic Challenges in Patients with Castleman Disease, a Single Center Experience from Hungary. *Pathology and Oncology Research*, **30**, Article ID: 1611785. https://doi.org/10.3389/pore.2024.1611785

[5] Wang, T., Chen, X., Chen, W., Shi, L. and Liu, J. (2022) A Retrospective Study of 44 Patients with Head and Neck Castleman's Disease. *European Archives of Oto-Rhino-Laryngology*, **279**, 2625-2630. https://doi.org/10.1007/s00405-021-07065-0

[6] Hoffmann, C., Oksenhendler, E., Littler, S., Grant, L., Kanhai, K. and Fajgenbaum, D.C. (2024) The Clinical Picture of Castleman Disease: A Systematic Review and Meta-Analysis. *Blood Advances*, **8**, 4924-4935. https://doi.org/10.1182/bloodadvances.2024013548

[7] Chen, Y., Zhang, W., Sun, C., OuYang, D., Chen, W., Luo, R., *et al.* (2012) Clinical Features and Outcomes of Head and Neck Castleman Disease. *Journal of Oral and*

Maxillofacial Surgery, **70**, 2466-2479. https://doi.org/10.1016/j.joms.2011.12.002

[8] Madan, R., Chen, J., Trotman-Dickenson, B., Jacobson, F. and Hunsaker, A. (2012) The Spectrum of Castleman's Disease: Mimics, Radiologic Pathologic Correlation and Role of Imaging in Patient Management. *European Journal of Radiology*, **81**, 123-131. https://doi.org/10.1016/j.ejrad.2010.06.018

[9] Pitot, M.A., Tahboub Amawi, A.D., Alexander, L.F., LeGout, J.D., Walkoff, L., Navin, P.J., *et al.* (2023) Imaging of Castleman Disease. *RadioGraphics*, **43**, e220210. https://doi.org/10.1148/rg.220210

[10] Cronin, D.M.P. and Warnke, R.A. (2009) Castleman Disease: An Update on Classification and the Spectrum of Associated Lesions. *Advances in Anatomic Pathology*, **16**, 236-246. https://doi.org/10.1097/pap.0b013e3181a9d4d3

[11] Dispenzieri, A. and Fajgenbaum, D.C. (2020) Overview of Castleman Disease. *Blood*, **135**, 1353-1364. https://doi.org/10.1182/blood.2019000931

[12] Lang, E. and van Rhee, F. (2024) Idiopathic Multicentric Castleman Disease: An Update in Diagnosis and Treatment Advances. *Blood Reviews*, **64**, Article ID: 101161. https://doi.org/10.1016/j.blre.2023.101161

[13] Talat, N., Belgaumkar, A.P. and Schulte, K. (2012) Surgery in Castleman's Disease: A Systematic Review of 404 Published Cases. *Annals of Surgery*, **255**, 677-684. https://doi.org/10.1097/sla.0b013e318249dcdc

[14] Jitaru, C., Zlampa, N., Dima, D., Bojan, A., Zdrenghea, M., Urian, L., *et al.* (2026) Snapshot Look at Castleman Disease. *Journal of Cellular and Molecular Medicine*, **30**, e70961. https://doi.org/10.1111/jcmm.70961