International Journal of Research in Medical Sciences
Dandala SR et al. Int J Res Med Sci. 2025 Jul;13(7):3027-3029
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: https://dx.doi.org/ 10.18203/2320-6012.ijrms20252043

Case Report

Unicentric hyaline vascular Castleman's disease presenting as an intraparenchymal parotid mass: a rare case report

Sridhar R. Dandala, Vinod K. Gonuru*, Manoranjani Dola

Department of Otorhinolaryngology, Apollo Institute of Medical Sciences and Research, Hyderabad, Telangana, India

Received: 26 March 2025
Revised: 01 May 2025
Accepted: 09 June 2025

***Correspondence:**
Dr. Vinod K. Gonuru,
E-mail: drvinnu2004@yahoo.co.in

ABSTRACT

Castleman's disease (CD) is a rare, benign lymphoproliferative disorder of unknown etiology that can affect any lymph node group, with the mediastinum being the most common site. Salivary gland involvement is exceptionally rare. We present a case of a 30-year-old female with a slowly progressive, painless left-sided parotid swelling persisting for four months. Clinical examination, ultrasonography and fine-needle aspiration cytology (FNAC) initially suggested an intraparotid lymph node enlargement. The lesion was excised and histopathological examination confirmed hyaline vascular CD. Given its rarity and lack of specific clinical or radiological features, CD should be considered in the differential diagnosis of persistent parotid gland swellings. Surgical excision remains the mainstay of treatment for unicentric Castleman's disease (UCD) with an excellent prognosis.

Keywords: Castleman's disease, Benign lymphoproliferative disorder, Parotid gland

INTRODUCTION

Castleman's disease (CD), first described by Dr. Benjamin Castleman in 1956, is an uncommon lymphoproliferative disorder with an unclear etiology.[1] CD can occur anywhere throughout the lymphatic system, most frequently in the mediastinum (60–86%), followed by the head and neck, whereas salivary gland involvement accounts for only 6–14% of cases.[2] CD has been referred to by multiple terms, including angiofollicular lymph node hyperplasia, angiofollicular hamartoma, angiomatous lymph node hamartoma, benign giant lymphoma and giant lymph node hyperplasia.[3] The disease manifests in two clinical forms: unicentric CD (UCD) and multicentric CD (MCD). The clinical manifestations and prognosis of UCD and MCD are usually different. UCD, which typically presents as an isolated lymph node enlargement without systemic symptoms, and MCD is aggressive and associated with systemic symptoms such as generalized lymphadenopathy, hepatosplenomegaly, fatigue, fever, sweat, arthralgia and weight loss.[4] Histologically, CD is classified into three types hyaline vascular (HV) type, plasma cell type and mixed type with the hyaline vascular subtype being the most common in head and neck cases (80–90%).[5,6] CD is in a group of very rare heterogeneous diseases, but it's exact etiology and pathogenesis remains unknown, although interleukin-6 (IL-6) is suspected to play a significant role in lymphoid proliferation.[7]

CASE REPORT

A 30-year-old female presented to the Department of Otorhinolaryngology with a painless, progressively enlarging swelling in the left parotid region over four months. The patient had no history of trauma, systemic illness or associated symptoms such as fever, fatigue, weight loss or malaise.

Clinical examination revealed a firm, non-tender, non-pulsatile, well-circumscribed 2×2 cm mass with a smooth surface and normal overlying skin. No signs of inflammation, palpable cervical lymphadenopathy or facial nerve involvement were noted.

Imaging and cytological findings

Ultrasonography (USG) of the left parotid gland showed a bulky gland with multiple well-defined hypoechoic lesions and increased vascularity on color Doppler, the largest measuring 16×11 mm, suggestive of necrotic intraparotid lymph nodes (Figure 1). Fine needle aspiration cytology (FNAC) from left parotid swelling- features are suggestive of chronic non-specific lymphadenitis.

Given the persistent nature of the lesion, an excision biopsy was performed (Figure 2).

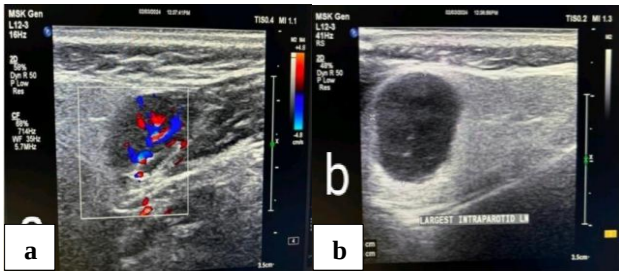

Figure 1 (a and b): Ultrasound images.

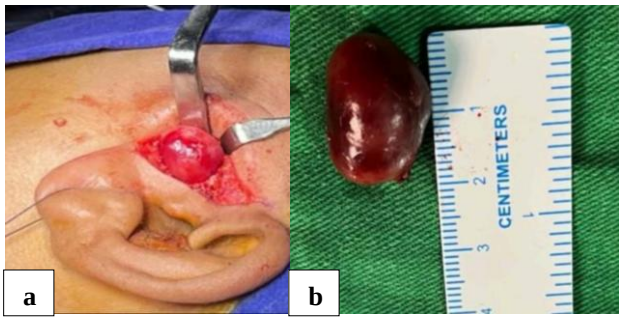

Figure 2 (a and b): Intraoperative view of intraparotid lymph node.

Treatment and follow up

Surgical excision remains the gold standard for UCD, offering excellent outcomes with a low recurrence rate. In contrast, MCD requires systemic therapy, including corticosteroids, monoclonal antibodies (e.g., siltuximab or tocilizumab) targeting IL-6 or chemotherapy (e.g. cyclophosphamide, vincristine, doxorubicin) in severe cases. The prognosis for UCD is excellent, with complete surgical excision leading to a curative outcome.[11] In the present case, the patient underwent complete excision of the lesion. The patient recovered well after the surgery and remained asymptomatic on follow-up.

Histopathological examination

Microscopic examination revealed a hyperplastic lymph node with numerous follicles of varying sizes, expanded germinal centers, and prominent hyalinized vessels traversing the follicles. The interfollicular areas contained plasma cells, histiocytes, and immunoblasts, confirming the diagnosis of hyaline vascular CD (Figure 3).

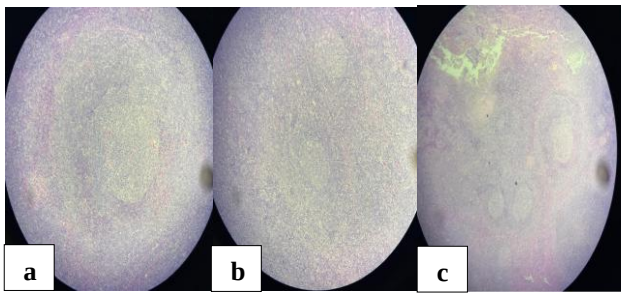

Figure 3 (a-c): Histopathological images.

DISCUSSION

CD is an uncommon and poorly understood lymphoproliferative disorder that may present across a wide age spectrum, from infancy to late adulthood, with a peak incidence in young adults. It exhibits no clear sex predilection. The hyaline vascular variant is most often associated with UCD, while the plasma cell variant is linked to MCD.[8] Mixed variant type is a rare variant and exhibits features of hyaline-vascular and plasma cell type .The mediastinum is the most frequently affected site (70%), while head and neck involvement is rare (15%).[9] Salivary gland involvement, particularly in the parotid gland is exceedingly rare, making this case an exceptional presentation.

Patients with UCD are usually asymptomatic, and their laboratory parameters are typically unremarkable. Conversely, MCD presents with systemic symptoms and may be associated with human herpesvirus-8 (HHV-8) or human immunodeficiency virus (HIV) infection.[10] IL-6 overexpression has been implicated in the pathogenesis of CD, particularly in MCD, where it contributes to systemic inflammation and lymphoid hyperplasia.

Due to its non-specific clinical presentation, CD is often misdiagnosed preoperatively. FNAC findings are frequently inconclusive, necessitating histopathological confirmation. Imaging modalities such as contrast-enhanced computed tomography (CT) or magnetic resonance imaging (MRI) may aid in diagnosis by identifying the vascular nature of the lesion, but definitive diagnosis is histopathological.

CONCLUSION

CD is a rare lymphoproliferative disorder with an elusive etiology, diverse clinical presentations, and histological subtypes. Salivary gland involvement, particularly in the parotid gland, is exceedingly rare, necessitating a high index of suspicion when evaluating persistent, painless parotid swellings. Diagnosis relies on histopathological examination, as FNAC and imaging findings are often inconclusive. Surgical excision remains the treatment of choice for UCD, offering an excellent prognosis. This case

underscores the importance of considering CD in the differential diagnosis of intraparotid lymphadenopathy and highlights the critical role of histopathology in definitive diagnosis.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Castleman B, Towne VW. Case records of the Massachusetts General Hospital; weekly clinicopathological exercises; Case No. 40351. N Engl J Med. 1954;251(10):396-400.
2. Batra A, Goyal M. Castleman's disease presenting as parotid mass: a case report. Int J Otorhinolaryngol Head Neck. 2019;5(2):504-6.
3. Gürbüzler L, Ceylan A, Yilmaz M, Vural C. Castleman's disease of the parotid gland: a case report. Kaohsiung J Med Sci. 2010;26(8):444-7.
4. Iyer S, Bhatti MI, Halliday M. Castleman's disease-A case report. Int J Surg Case Rep. 2010;1(3):25-6.
5. Keller AR, Hochholzer L, Castleman B. Hyaline-vascular and plasma-cell types of giant lymph node hyperplasia. Cancer. 1972;29(3):670-83.
6. Samadi, Daniel S, Hockstein NG, Tom LWC. Pediatric intraparotid Castleman's disease. Ann Otol Rhinol Laryngol. 2003;112(9):813-6.
7. Leger-Ravet MB, Peuchmaur M, Devergne O, Audouin J, Raphael M, Van Damme J, et al. Interleukin-6 gene expression in Castleman's disease. Blood. 1991;78(11):2923-30.
8. Haap M, Wiefels J, Horger M, Hoyer A, Müssig K. Clinical, laboratory and imaging findings in Castleman's disease - The subtype decides. Blood Rev. 2018;32(3):225-34.
9. Williams DS. Castleman's disease. J Insur Med. 2013;43(4):256-8.
10. Reece B, Ord R, Papadimitriou J. Rare presentation of unicentric Castleman's disease in the parotid gland. J Oral Maxillofac Surg. 2012;70(9):2114-7.
11. Bucher P, Chassot G, Zufferey G, Ris F, Huber O, Morel P. Surgical management of abdominal and retroperitoneal Castleman's disease. World J Surg Oncol. 2005;3:33.

Cite this article as: Dandala SR, Gonuru VK, Dola M. Unicentric hyaline vascular Castleman's disease presenting as an intraparenchymal parotid mass: a rare case report. Int J Res Med Sci 2025;13:3027-9.