Received: 22 August 2023 | Revised: 7 November 2023 | Accepted: 10 November 2023

DOI: 10.1002/ccr3.8258

CASE REPORT

Clinical Case Reports Open Access WILEY

Unicentric Castleman disease presenting as a longstanding axillary and chest wall mass: A case report

Wenqing Zhou[1] | **Xing Liu**[2] | **Aiming Qiu**[3] | **Teng Ni**[1] | **Tiangeng Dong**[4] | **Lei Ding**[5]

[1]Department of General Surgery, The Fifth People's Hospital of Wujiang Area, Suzhou, Jiangsu, China

[2]Department of Pain Treatment, The Fifth People's Hospital of Wujiang Area, Suzhou, Jiangsu, China

[3]Department of General Practice, The Fifth People's Hospital of Wujiang Area, Suzhou, Jiangsu, China

[4]Department of General Surgery, Zhongshan Hospital, Fudan University, Shanghai, China

[5]Department of Pediatrics, The Fifth People's Hospital of Wujiang Area, Suzhou, Jiangsu, China

Correspondence

Lei Ding, Department of Pediatrics, The Fifth People's Hospital of Wujiang Area, Suzhou, Jiangsu 215200, China.
Email: dingleiokok@163.com

Tiangeng Dong, Department of General Surgery, Zhongshan Hospital, Fudan University, Shanghai, China.
Email: dong.tiangeng@zs-hospital.sh.cn

Funding information

the Science and Education Foundation of Wujiang District, Grant/Award Number: wwk202019

Key clinical message

Unicentric Castleman disease, particularly the hypervascular variant subtype, commonly presents as a localized lymphadenopathy without systemic symptoms. Surgical excision is often curative for this subtype, leading to a good prognosis. However, some patients with autoimmune complications may require additional systemic therapy along with surgery. Accurate diagnosis through a combination of clinical, radiological, and pathological findings is crucial for optimal management.

KEYWORDS

axillary, chest wall, unicentric Castleman disease

1 | INTRODUCTION

Castleman disease, first described in 1956 by Dr. Benjamin Castleman, is a rare lymphoproliferative disorder. This disease can affect any lymphoid tissue in the body and present with localized or systemic symptoms. The diagnosis is based on pathological examination, and the disease has a varied clinical course and response to treatment.[1]

2 | CASE REPORT

The patient was a 30-year-old female with a ten-year history of a progressively enlarging mass in the left axilla and chest wall (Figure 1). The mass was initially small and asymptomatic but had gradually increased in size over the years. The patient had no significant medical history or family history of lymphoproliferative disorders.

Wenqing Zhou, Xing Liu, and Aiming Qiu contributed equally to this work.

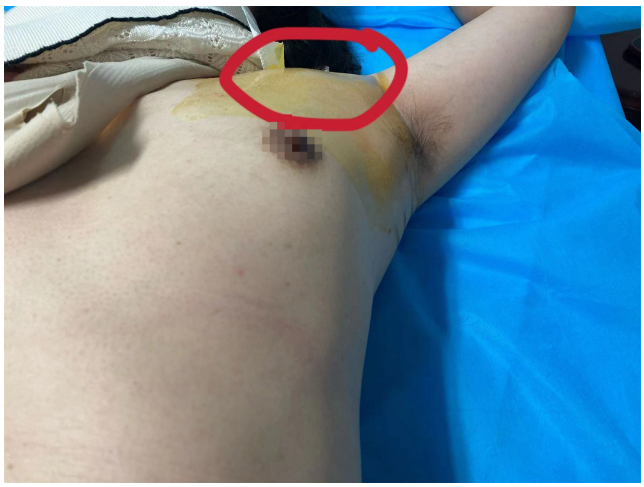

FIGURE 1 Location of the mass is significantly deeper than the skin and visible to the naked eye.

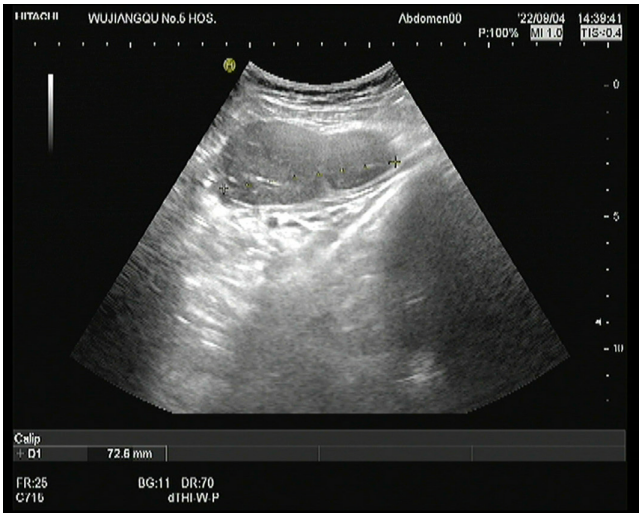

FIGURE 2 Ultrasound examination revealed an intact tumor capsule, presenting as an irregular hypoechoic mass.

Diagnostic workup, including ultrasonography (Figure 2) and MRI [Figure 3], revealed a well-circumscribed, heterogeneously enhancing mass measuring 10 × 6 × 3 cm. The laboratory investigations revealed that the patient's white blood cell count and red blood cell counts were within normal range. Additionally, the CRP (C-reactive protein) and ESR (erythrocyte sedimentation rate) results were also normal. Notably, the IgG level was measured to be 10.6 g/L. A core needle biopsy was performed, and pathological examination revealed follicular hyperplasia, onion skin-like changes, increased vascularity in the interfollicular areas, and proliferation of fibrous tissue in the capsule (Figure 4). The histopathological features were consistent with the hyaline vascular variant of Castleman disease.[2]

After appropriate preoperative preparation, the patient underwent surgical excision of the mass. Intraoperative findings revealed a well-encapsulated mass that was adherent to the chest wall muscles. The mass was completely excised with negative margins. Postoperative recovery was uneventful, and the patient was discharged on postoperative Day 5.

Histopathological examination of the excised mass confirmed the diagnosis of Castleman disease. HE staining revealed follicular hyperplasia, onion skin-like changes, increased vascularity in the interfollicular areas, and proliferation of fibrous tissue in the capsule. These findings are consistent with Castleman disease. The diagnosis of unicentric Castleman disease was determined based on the patient's medical history, physical examination, medical diagnostic tests, surgery, and pathological examination.

3 | DISCUSSION

Castleman disease is a rare lymphoproliferative disorder that can present with varied clinical features.[3] Castleman disease is first divided into unicentric and multicentric, and multicentric CD is further subdivided into POEMS-associated, HHV-8+, or HHV-8- (idiopathic multicentric Castleman disease). HHV-8-iMCD is further divided into iMCD-TAFRO and iMCD-NOS.[4] The disease has two major histological subtypes: hyaline vascular (or Hypervascular variant) and plasma cell. The more common hypervascular variant subtype presents with localized lymphadenopathy, and this is the histology most commonly associated with unicentric Castleman disease,[5] while the less common plasma cell subtype is associated with systemic symptoms and multiorgan involvement.[6] However, in this subset of iMCD-TAFRO patients, there are also individuals who exhibit the hypervascular variant subtype, which is the most aggressive form of CD and has severe systemic symptoms. In this case study, a patient with unicentric Castleman disease was confirmed to have the Hypervascular variant subtype based on pathological examination. The patient underwent complete surgical resection without receiving any additional systemic treatment. Following a one-year postoperative follow-up, no signs of recurrence or complications were observed. In general, for unicentric Castleman disease, surgical resection is considered curative, but some patients with unicentric CD have autoimmune complications such as immune-mediated cytopenias and bullous pemphigus, and these patients have a more aggressive course, often requiring systemic therapy in addition to surgery.[7]

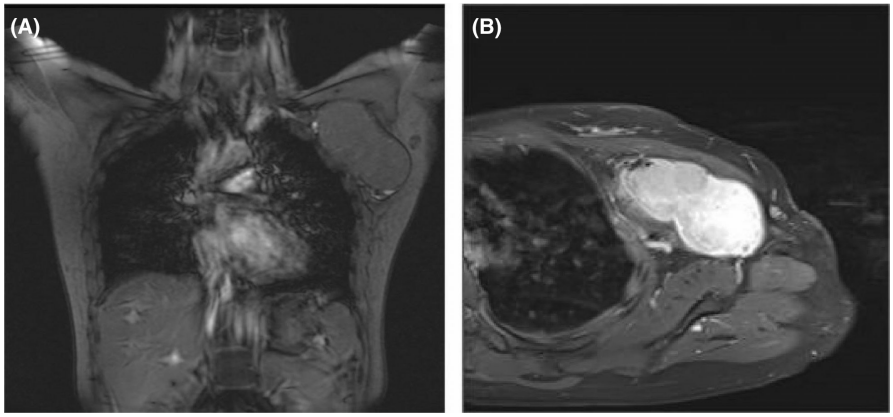

FIGURE 3 Magnetic resonance imaging (MRI) exhibited well-defined lesion borders with a larger cross-sectional size of approximately 99 mm * 53 mm. The T1-weighted images demonstrated isointense signal, while the T2-weighted images showed slightly hyperintense signal. The presence of flow voids consistent with blood vessels was observed within the lesion. Following contrast administration, there was a significant and homogeneous enhancement.

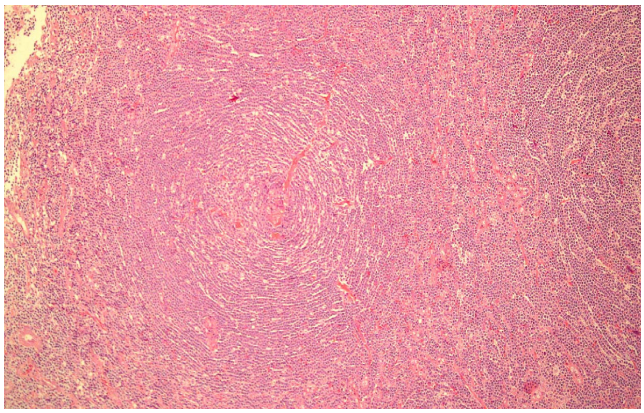

FIGURE 4 HE staining revealed follicular hyperplasia, onion skin-like changes, increased vascularity in the interfollicular areas, and proliferation of fibrous tissue in the capsule. These findings are consistent with Castleman disease.

Diagnosis of Castleman disease requires a combination of clinical, radiological, and pathological findings. Treatment options include surgery, radiation therapy, and chemotherapy, depending on the subtype, stage, and extent of the disease.[8] The localized hypervascular variant subtype generally has a good prognosis, while the systemic plasma cell subtype has a more variable clinical course.[9]

4 | CONCLUSION

Castleman disease should be considered as a differential diagnosis for longstanding masses in lymphoid tissue. Appropriate diagnostic workup, including imaging and pathological examinations, is crucial for accurate diagnosis and optimal management. Surgical excision can offer a curative treatment with a good prognosis for the localized hyaline vascular subtype.[10]

AUTHOR CONTRIBUTIONS

Wenqing Zhou: Writing – original draft. **Xing Liu:** Investigation. **Aiming Qiu:** Investigation. **Teng Ni:** Data curation. **Tiangeng Dong:** Resources; writing – review and editing. **Lei Ding:** Writing – review and editing.

FUNDING INFORMATION

This work was supported by grants from the Science and Education Foundation of Wujiang District (wwk202019).

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

ORCID

Wenqing Zhou https://orcid.org/0000-0002-0974-591X

REFERENCES

1. Oksenhendler E. The Spectrum of Castleman's disease and its treatment. *Hematology Am Soc Hematol Educ Program*. 2016;2016(1):327-334.
2. Chan KL, Tong J, Loong F, et al. Castleman's disease: a report of 18 cases from a single institution. *J Clin Oncol*. 1998;16(5):1988-1995.
3. Soumerai JD, Sohani AR, Abramson JS. Diagnosis and management of Castleman disease. *Cancer Control*. 2014;21(4):266-278.

4. Chen L, Fajgenbaum DC. Castleman disease. In: Stone JH, ed. *A Clinician's Pearls & Myths in Rheumatology*. Springer International Publishing; 2023.
5. Dispenzieri A, Fajgenbaum DC. Overview of Castleman disease. *Blood*. 2020;135(16):1353-1364.
6. Dispenzieri A. Castleman disease. *Hematology Am Soc Hematol Educ Program*. 2018;2018(1):324-329. doi:10.1182/asheducation-2018.1.324
7. Dieudonne Y, Silvestrini MA, Dossier A, et al. Paraneoplastic pemphigus uncovers distinct clinical and biological phenotypes of western unicentric Castleman disease. *Br J Haematol*. 2023;202(2):267-278.
8. Liu AY, Nabel CS, Finkelman BS, et al. Idiopathic multicentric Castleman's disease: a systematic literature review. *Lancet Haematol*. 2016;3(4):e163-e175. doi:10.1016/S2352-3026(16)00015-X
9. Talat N, Belgaumkar AP, Schulte KM. Surgery in Castleman's disease: a systematic review of 404 published cases. *Ann Surg*. 2012;255(4):677-684. doi:10.1097/SLA.0b013e31824a57e9
10. van Rhee F, Stone K, Szmania S, Barlogie B, Singh Z. Castleman disease in the 21st century: an update on diagnosis, assessment, and therapy. *Clin Adv Hematol Oncol*. 2010;8(7):486-498.

How to cite this article: Zhou W, Liu X, Qiu A, Ni T, Dong T, Ding L. Unicentric Castleman disease presenting as a longstanding axillary and chest wall mass: A case report. *Clin Case Rep*. 2023;11:e8258. doi:10.1002/ccr3.8258