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Case Report

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Symptomatic unicentric Castleman disease with hilar localization mimicking malignancy: a diagnostic challenge

Ayşen Atasoy[1], Esma Sevil Akkurt[1], Özlem Düvenci Birben[1], Muhammet Ali Beyoğlu[2], Derya Yenibertiz[1]

[1]*Department of Pulmonology, Dr. Abdurrahman Yurtaslan Ankara Oncology Training and Research Hospital, University of Health Sciences, Ankara, Turkiye*
[2]*Department of Thoracic Surgery, Dr. Abdurrahman Yurtaslan Ankara Oncology Training and Research Hospital, University of Health Sciences, Ankara, Turkiye*

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Corresponding Author: Esma Sevil Akkurt, esma.sevil@hotmail.com

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ABSTRACT

Unicentric Castleman disease (UCD) is a rare lymphoproliferative disorder that is often detected incidentally and asymptomatically. However, in some cases, mass effect or inflammatory features may cause clinical symptoms, posing a diagnostic challenge. A 43-year-old woman with a history of toxic nodular goiter presented with dyspnea. Thoracic CT revealed a 37×29 mm well-circumscribed mass in the left hilum encasing the basal branch of the pulmonary artery. PET-CT showed moderate FDG uptake (SUVmax: 6.1). EBUS-guided biopsy reported reactive lymphoid tissue. Due to diagnostic uncertainty, surgical excision was performed. Histopathology confirmed hyaline-vascular type UCD. HHV-8-associated MCD and lymphoma were excluded. This case highlights the diagnostic complexity of UCD, especially when presenting with a symptomatic hilar mass and moderate FDG uptake mimicking malignancy. Surgical biopsy remains essential for definitive diagnosis.

Keywords: Unicentric Castleman disease, hyaline-vascular type, hilar localization; FDG-PET, excisional biopsy

INTRODUCTION

Castleman disease (CD), first described by Castleman et al.[1] in 1954, is a rare lymphoproliferative disorder characterized by lymph node hyperplasia. Clinically, it is classified into two main forms: unicentric Castleman disease (UCD) and multicentric Castleman disease (MCD).[2,3] UCD typically involves a single lymph node or nodal station and is often asymptomatic.[3] Diagnosis is usually made incidentally during imaging studies performed for other reasons. However, in rare cases, clinical symptoms may develop due to the mass effect of the lesion.[4] While the mediastinum is the most frequently reported site of involvement, hilar localization remains exceedingly rare.[5,6]

The estimated annual incidence of CD is approximately 21-25 cases per million population, with unicentric forms accounting for the majority.[2,9] A definitive diagnosis of UCD can only be established by histopathological examination, as minimally invasive biopsies are often inconclusive.[6,7] In this context, symptomatic UCD with hilar localization is exceedingly rare and may mimic malignancy during the diagnostic process. Herein, we present a case of hilar UCD encasing the pulmonary artery with moderate fluorodeoxyglucose (FDG) uptake on positron emission tomography–computed tomography (PET-CT), histopathologically diagnosed as the hyaline-vascular subtype, highlighting the diagnostic challenges and surgical management.

CASE

A 43-year-old female patient presented to the pulmonology outpatient clinic with complaints of shortness of breath. Her only known medical condition was toxic nodular goiter. She was a lifelong non-smoker. Physical examination findings were unremarkable, and pulmonary function tests were within normal limits. Laboratory tests revealed anemia, leukocytosis, and elevated C-reactive protein (CRP) levels, although there were no clinical signs of infection.

A chest X-ray showed fullness in the left hilar region. Thoracic computed tomography (CT) revealed a well-circumscribed mass measuring 37×29 mm in the left hilum, encircling 180 degrees of the basal branch of the lower lobe pulmonary artery, along with a 15×11 mm lymphadenopathy in the subcarinal area (**Figure 1**).

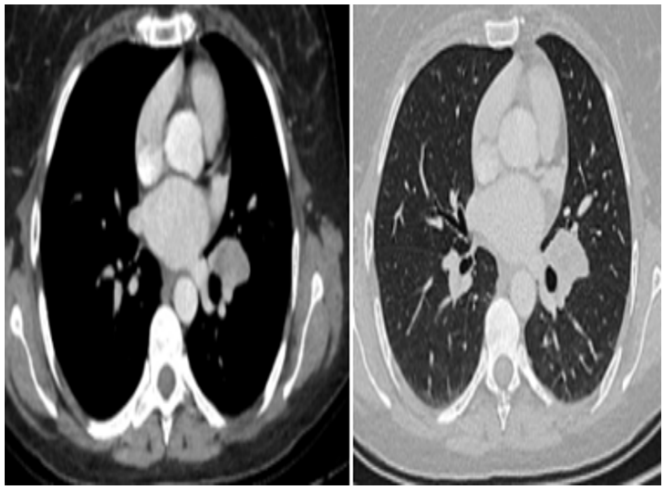

Figure 1. Thoracic computed tomography (CT) showing a well-circumscribed mass (37×29 mm) in the left hilum encircling the basal branch of the lower lobe pulmonary artery, with a 15×11 mm subcarinal lymph node

On PET-CT, a mass-like lesion of approximately 4 cm in the left hilar region demonstrated moderate FDG uptake (SUVmax: 6.1) (**Figure 2**). The differential diagnosis included tuberculosis, sarcoidosis, and lymphoma. Sampling of subcarinal and left infrahilar lymph nodes was performed via endobronchial ultrasound (EBUS). Cytological examination was consistent with a reactive lymph node pattern.

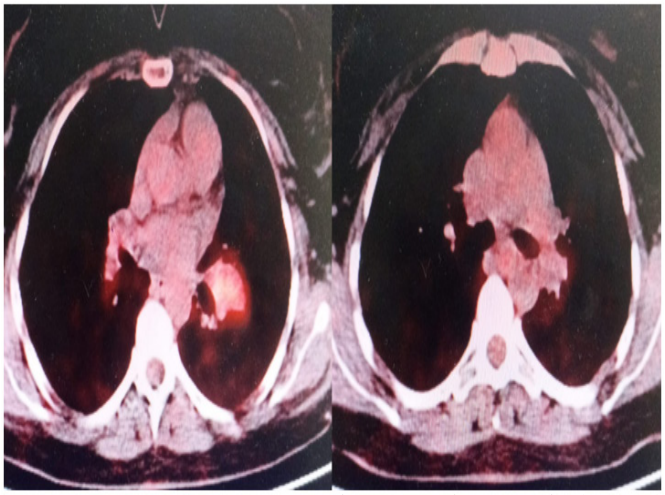

Figure 2. Positron emission tomography–computed tomography (PET-CT) demonstrating moderate fluorodeoxyglucose (FDG) uptake (SUVmax=6.1) in the left hilar mass

As the diagnosis remained inconclusive, the patient was referred to thoracic surgery, and an excisional biopsy was planned. Following surgical excision, histopathological examination confirmed the diagnosis of hyaline-vascular type CD (**Figure 3**). To exclude human herpesvirus 8 (HHV-8)–associated MCD and lymphoma, serum protein electrophoresis, immunofixation, Epstein–Barr virus (EBV) PCR, 24-hour urine protein analysis, and urine electrophoresis were performed, all of which were within normal limits. Beta-2 microglobulin level was 2.83 mg/L, and lactate dehydrogenase (LDH) was 161 U/L. Although both values were normal, elevations in these parameters may be observed in multicentric forms, helping to distinguish them from UCD. The patient was followed postoperatively without complications.

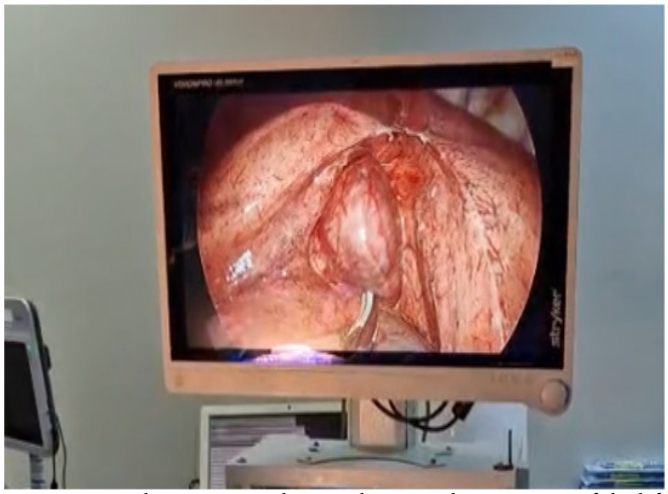

Figure 3. Bronchoscopic view showing the external compression of the left lower lobe bronchus caused by a well-circumscribed hilar mass

DISCUSSION

UCD is a rare lymphoproliferative disorder that is often asymptomatic and usually detected incidentally during radiological examinations performed for unrelated reasons.[2,3] While the mediastinum is the most common site of involvement, UCD has also been reported in various anatomical regions, such as the neck, mesentery, and retroperitoneum.[5,6] Hilar localization, particularly on the left side, is extremely rare. Only a few cases of hilar UCD have been reported in the literature, and these cases similarly presented with nonspecific respiratory symptoms and imaging findings mimicking malignancy.

Although UCD is typically asymptomatic, some patients may develop localized symptoms, such as dyspnea, chest pain, or cough, due to the mass effect of the lesion.[4,8] In our patient, dyspnea was likely related to the mass encasing the pulmonary vasculature. Laboratory findings, including leukocytosis, anemia, and elevated CRP, resembled systemic inflammatory features more commonly associated with MCD.[2,9] However, mild inflammatory responses have also been reported in some UCD cases.[9] Normal beta-2 microglobulin and LDH levels in our patient further supported the unicentric form, as elevations in these parameters are more suggestive of multicentric disease.

One of the major diagnostic challenges in our case was the presence of imaging features suggestive of malignancy. PET-CT demonstrated moderate FDG uptake (SUVmax: 6.1) within the lesion, raising suspicion for malignancy. Although PET-CT is useful for identifying metabolically active lesions, its specificity in differentiating benign from malignant conditions is limited.[10,11] Therefore, PET-CT findings alone are insufficient for definitive diagnosis.

EBUS-guided biopsy in our patient revealed only reactive lymphoid tissue, which was inadequate for diagnosis. As reported in the literature, minimally invasive biopsies have limited diagnostic yield in UCD, and excisional biopsy remains the gold standard.[6,7] In our patient, histopathological

examination following surgical excision confirmed the diagnosis of hyaline-vascular type UCD. This subtype is the most common in UCD and is associated with a favorable prognosis and low recurrence rate.[2,12] Complete surgical resection in UCD is associated with excellent long-term survival and minimal risk of recurrence.[6] In our case, total excision was achieved, the postoperative period was uneventful, and the patient remains stable under follow-up.

CONCLUSION

A left hilar mass encasing the pulmonary artery with moderate FDG uptake initially raised suspicion for malignancy; however, the definitive diagnosis of hyaline-vascular type UCD was established only after surgical excision. CD, particularly in uncommon locations, can clinically and radiologically mimic malignancy. In symptomatic cases with hilar localization and FDG uptake, the differential diagnosis should remain broad, and surgical excision should be strongly considered when minimally invasive methods are inconclusive, as it remains the gold standard for definitive diagnosis.

ETHICAL DECLARATIONS

Informed Consent

Written informed consent has been obtained from the patient included in this report. Signed consent forms are retained by the authors and are available upon request.

Peer Review Process

This report underwent external peer review.

Conflict of Interest

The authors declare no conflicts of interest.

Financial Disclosure

This case report did not receive any financial support.

Author Contributions

All authors made substantial contributions to the clinical documentation, interpretation, and manuscript preparation. All authors approved the final version of the manuscript.

REFERENCES

1. Castleman B, Towne VW. Case records of the Massachusetts General Hospital: Case No. 40231. *N Engl J Med.* 1954;250(23):1001-1005. doi:10.1056/NEJM195406102502308
2. Fajgenbaum DC, Uldrick TS, Bagg A, et al. International, evidence-based consensus diagnostic criteria for HHV-8–negative/idiopathic multicentric Castleman disease. *Blood.* 2017;129(12):1646-1657. doi:10.1182/blood-2016-10-746933
3. UpToDate: "Unicentric Castleman disease: clinical features, evaluation, and diagnosis." Erişim: July 2025.
4. Harisinghani MG, Kalra MK, Dineen S, et al. Radiologic features of unicentric Castleman disease on CT. *Clin Radiol.* 2012;67(2):137-143.
5. Bonekamp D, Horton KM, Hruban RH, Fishman EK. Castleman disease: the great mimic. *Radiographics.* 2011;31(6):1793-1807. doi:10.1148/rg.316115502
6. 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.0b013e318249dcdc
7. Bowne WB, Lewis JJ, Filippa DA, et al. The management of unicentric and multicentric Castleman's disease: a report of 16 cases and a review of the literature. *Cancer.* 1999;85(3):706-717. doi:10.1002/(sici)1097-0142(19990201)85:3<706::aid-cncr21>3.0.co;2-7
8. Frizzera G. Castleman's disease and related disorders. *Semin Diagn Pathol.* 1988;5(4):346-364.
9. 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)00006-5
10. Kato Y, Kojima M, Yatsui SK. Usefulness of FDG-PET in differentiating Castleman's disease from lymphoma. *J Nucl Med.* 2018;59(suppl 1):19.
11. Ueno T, Suzui N, Tamura A, et al. FDG-PET/CT for evaluation of Castleman disease. *Cancer Imaging.* 2019;19(1):25.
12. Chen D-Y, Chen H-Y, Hou C-H, et al. Clinicopathological features and prognosis of hyaline vascular subtype unicentric Castleman disease. *J Thorac Dis.* 2020;12(12):7628-7636.