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# CASTLEMAN DISEASE: A CASE REPORT

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**ABSTRACT**

Castleman disease (CD) is a group of lymphoproliferative disorders with common lymph node histological features. It may be either unicentric or multicentric. Unicentric Castleman disease (UCD) is localized and carries an excellent prognosis, whereas multicentric Castleman disease (MCD) is a systemic disease occurring most commonly in the setting of HIV infection and is associated with human herpesvirus 8.[1]Castleman disease in the abdomen and pelvis is rare and liable to misdiagnosis, but its characteristic imaging features can help in the diagnosis and differential diagnosis.[2]Unicentric Castleman disease (UCD) is rare, and there are no reliable estimates of its incidence in the population. While UCD can occur at any age, it is generally a disease of younger adults. The median age at presentation is approximately 35 years.[3, 4]

***Keywords***: *Castleman Disease; Infections; Lymph Nodes;*

## CASE PRESENTATION

A 13-year-old boy presented to the medicine outpatient department for the evaluation of fever, anorexia, and weight loss without any history of cough. Physical examination showed few right cervical lymphadenopathies. No organomegaly or tenderness was seen. Complete blood count and biochemical profile were within the normal limits. He was referred to the radiology department for computerized tomography (CT) scan of the abdomen which showed multiple discrete retroperitoneal and mesenteric lymphadenopathy (Figure 1).

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Variable-sized lymph nodes in the peripancreatic region showed homogenous lymph node enhancement in the early arterial phase of contrast study, largest measuring 3.4 cm in short axis. There was no hepatomegaly or splenomegaly. Chest X-ray was unremarkable. Few precarinal and aortocaval lymphadenopathy was also noted.

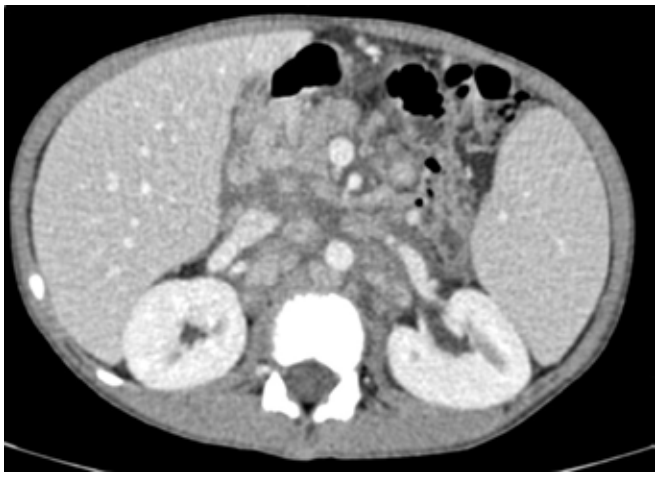

**Figure 1:** Contrast-enhanced computerized tomography (CECT) scan of the abdomen showing multiple discrete retroperitoneal and mesenteric lymphadenopathy.

Biopsy of palpable cervical lymph node was done. Section showed lymph nodes with thickened traversing fibrous septa and mostly preserved architecture (Figure 2). The lymph nodes consisted of lymphoid follicles located in the cortex, paracortex, and medulla which had a variable-sized germinal center with occasional twinning. Hyaline deposits were noted within a few germinal centers. There was a concentric arrangement of mature lymphocytes around the germinal center.

Interfollicular areas are composed of eosinophils, plasma cells, and plasmacytoid monocytes. There were sclerotic blood vessels as well in the interfollicular region. A final diagnosis of Castleman disease (Hyaline variant) was made.

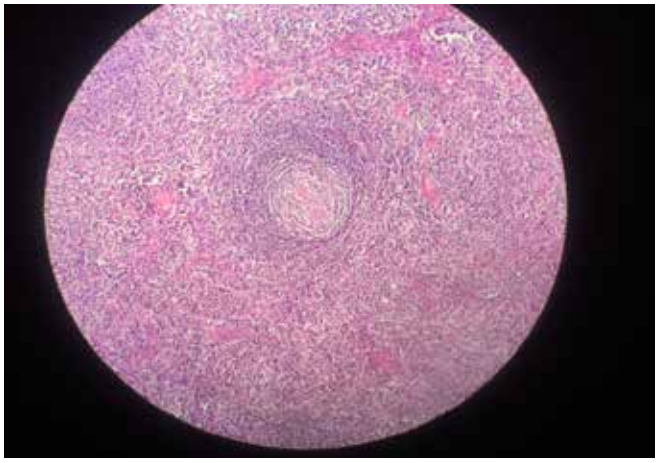

**Figure 2:** The follicle is surrounded by the broad mantle zone composed of small lymphocytes (onion skin) with a sclerosed germinal center.

## CASE DISCUSSION

Castleman disease is a rare, generally benign disease characterized by atypical proliferation of lymphocytes.[5] It is characterized as either unicentric or multicentric. Unicentric Castleman disease (UCD) is localized and carries an excellent prognosis, whereas multicentric Castleman disease (MCD) is a systemic disease occurring most commonly in the setting of HIV infection and is associated with human herpesvirus 8.[1] The most common site is the mediastinum (approximately 70%). Additional sites of occurrence include the axilla, retroperitoneum, mesentery, vulva, pancreas, pelvis, and neck.[6] Major histologic subtypes of Castleman disease are Hyaline vascular and Plasma cell types. Uncommonly, a transitional or mixed form may be seen. The hyaline vascular subtype is seen in 90% of cases, and 70% of cases are seen in patients younger than 30 years old.[7] Ultrasonographic findings are not specific and may resemble findings of lymphoma.[8]

Both hyaline-vascular and plasma cell types present as hypoechoic masses and nodules. In addition to vascular resistance assessed traditionally with Doppler spectral analysis, vascular pattern and vascular density assessed with power Doppler

sonography can better differentiate the nature of lymphadenopathies.

In a study, both metastatic lymphadenopathy and lymphoma showed high vascularity, probably as a result of tumor angiogenesis. Vascularity index is a more accurate criterion than Doppler spectral analysis. However, no matter which parameter was used for assessment, remarkable heterogeneity existed within each lymphadenopathy group.[9]

Common CT appearance is single, well-defined enhancing mass. Smaller tumors (<5 cm) display homogeneous contrast enhancement; larger tumors (>5 cm) show heterogeneous enhancement and attenuation when correlated with central necrosis and degeneration.[5] Obvious enhancement in the arterial phase and continuous enhancement in the portal vein and delayed phase is seen with an attenuation pattern similar to that of large vessels and enlarged blood vessels within or around the mass.[2]

The differential diagnosis which needs consideration

- Lymphoma: Non-Hodgkin lymphoma subtypes: Follicular lymphoma, Mantle cell lymphoma, and Nodal marginal zone B cell lymphoma. On computed tomographic (CT) images, involved lymph nodes are generally enlarged and of homogeneous density. Some lymphoma subtypes, namely small cell lymphocytic lymphoma/chronic lymphocytic leukemia, may manifest as an increased number of small nodes.[10]
- Follicular hyperplasia
- Toxoplasma lymphadenitis
- HIV lymphadenitis
- Metastasis

## CONCLUSION

There are multiple differential diagnoses to the enhancing lymph nodes in the abdomen. Castleman disease may be missed due to its rare incidence. It should be considered if a convincing enhancement pattern is observed in contrast-enhanced CT scan and definitive histological diagnosis should be obtained.

### CONFLICT OF INTEREST

None

### SOURCES OF FUNDING

None

## REFERENCES

1. Soumerai JD, Sohani AR, Abramson JS. Diagnosis and management of Castleman disease. *Cancer Control*. 2014;21(4):266-78. https://doi.org/10.1177/107327481402100403

2. Xiao L, Zhang ZL, Liu YB, et al. CT and MRI features of Castleman's disease of the abdomen and pelvis. *Nan Fang Yi Ke Da XueXueBao*. 2011;31(1):129-32.

3. Keller A, Hochholzer L, Castleman B. Hyaline-vascular and plasma-cell types of giant lymph node hyperplasia of the mediastinum and other locations. *Cancer*. 1972;29(3):670-83.https://doi.org/10.1002/1097-0142(197203)29:3<670::AID-CNCR2820290321>3.0.CO;2-#

4. Talat N, Belgaumkar AP, Schulte K-M. Surgery in Castleman's disease: a systematic review of 404 published cases. *Ann Surg*. 2012;255(4):677-684.https://doi.org/10.1097/SLA.0b013e318249dcdc

5. Meador TL, Mclarney JK. CT features of Castleman disease of the abdomen and pelvis. *Am J Roentgenol*. 2000;175(1):115-8. https://

doi.org/10.2214/ajr.175.1.1750115

6. Shin JH, Lee HK, Kim SY, Khang SK, Park SH, Choi CG, et al. Castleman's disease in the retropharyngeal space: CT and MR imaging findings. *Am J Neuroradiol*. 2000;21(7):1337–9

7. Keller A, Hochholzer L, Castleman B. Hyaline-vascular and plasma-cell types of giant lymph node hyperplasia of the mediastinum and other locations. *Cancer*. 1972;29(3):670-83. https://doi.org/10.1002/1097-0142(197203)29:3<670::AID-CNCR2820290321>3.0.CO;2-#

8. Williams JL, Kaude JV. Sonographic findings in a case of Castleman disease of the neck. *J Ultrasound Med*. 1986;5(10):593–4. https://doi.org/10.7863/jum.1986.5.10.593

9. Wu CH, Chang YL, Hsu WC, Ko JY, Sheen TS, Hsieh FJ. Usefulness of Doppler spectral analysis and power Doppler sonography in the differentiation of cervical lymphadenopathies. *Am J Roentgenol*.1998;171(2):503–9. https://doi.org/10.2214/ajr.171.2.9694484

10. Johnson SA, Kumar A, Matasar MJ, Schöder H, Rademaker J. Imaging for staging and response assessment in lymphoma. *Radiology*. 2015;276(2):323-38. https://doi.org/10.1148/radiol.2015142088