

Case report

Unicentric Castleman's disease presenting as a neck swelling: a case report

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ABSTRACT

Castleman's disease is an uncommon entity which is characterized by lymphadenopathy either at a single focus or at multiple sites. Due to the similar clinical picture of other disorders including tuberculous lymphadenitis, lymphoma and other cases of generalized lymphadenopathy like AIDS a diagnosis of castleman's disease. The unicentric form may resemble any generalized swelling such as a lipoma. In this case we discussed an 18 yr old girl presenting with a large neck swelling in the posterior triangle which clinically resembled a lipoma. The CT showed a uniformly enhancing homogenous lesion. After an excision biopsy, the histopathology revealed it to be castleman's disease of the hyaline vascular type. The literature reveals that although rare castleman's disease can be unicentric or multicentric and can involve multiple sites including the axilla, mediastinum and retroperitoneum. Most cases of the unicentric disease are completely cured with an excision biopsy. The multicentric disease may need chemotherapy, radiation or immunomodulating drugs. In conclusion, one must keep a differential of castleman's disease in mind when presented with any case of lymphadenopathy.

Keywords: Castleman's disease, Unicentric, Lymphadenopathy

INTRODUCTION

Castleman's disease was first described by Benjamin Castleman, after reporting 13 cases of mediastinal lymph node enlargement thought to be thymoma.¹ The majority of cases have been reported in the mediastinum, though other sites have also been noted in the literature.² It is an uncommon disease of benign etiology resulting in lymph node enlargement. Here, we discuss a case of an 18 year old girl presenting with a neck mass eventually diagnosed to be Castleman's disease.

CASE REPORT

An 18 year old girl presented to the OP with a painless mass in the right side of the neck which was initially small in size and gradually progressed over the span of 2

months to the present size. The swelling was noted in the posterior triangle of the right side of the neck measuring 7×4 cm in size, surface was smooth with a firm consistency. The swelling was mobile with no obvious cough impulse. Transillumination couldn't be appreciated. Clinically, a provisional diagnosis of lipoma was made.

Basic blood investigations revealed no significant abnormality. A contrast CT showed a homogenous, enhancing lesion in the posterior triangle of size 7×4 cm. Differential diagnosis included a vascular lesion, lymphadenopathy secondary to tuberculosis as well as lymphoma was considered. No infiltration into the surrounding tissue was noted. Hence an excision biopsy was planned.



Figure 1: Patient clinical photo.

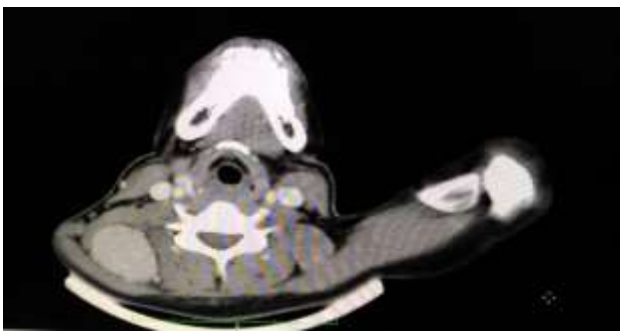


Figure 2: CECT(axial section).

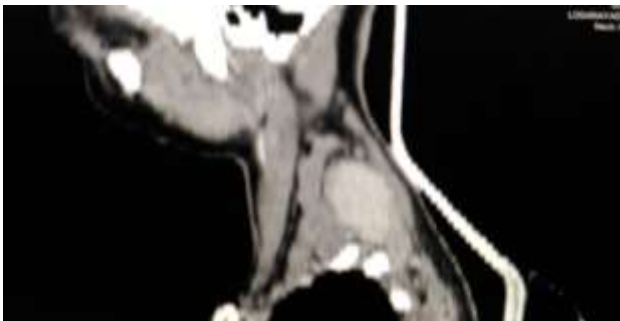


Figure 3: CECT (sagittal section).



Figure 4: Intra-operative photo.

Intra-operatively a large 6.5×3.7 cm swelling was excised. The swelling was below the plane of the muscle and had multiple feeding vessels which were serially ligated and finally excision done. Histopathology revealed, Castleman's disease of the hyaline vascular type.

DISCUSSION

Castleman's disease is a rare lymphoproliferative disorder of unknown etiology. It is also referred to as angiofollicular lymph node hyperplasia, giant lymph node hyperplasia, angiofollicular hamartoma, and benign lymphoma.² It is clinically divided into a localized unicentric (as in this case) or a multicentric variety. Histological classification includes the hyaline vascular, plasma cell, and mixed type. The unicentric form is mostly the hyaline vascular type and the multicentric form is mostly either the plasma cell or mixed type.³

The unicentric form of castleman disease most frequently manifests as an asymptomatic, solitary mediastinal tumor (40-70% of cases).^{4,5} Additional sites of occurrence include the axilla, retroperitoneum, mesentery, vulva, pancreas, pelvis, and neck.⁵ Less than 10% of cases occur in the neck.⁶

The multicentric form is rarer, with an increased incidence of systemic manifestations (including fever, malaise and anemia as well as hypergammaglobulinemia), along with a worse prognosis.⁷ A correlation with human immunodeficiency virus and human herpes virus 8 was noted. The postulated theory is due to dysregulation of interleukin-6. The multicentric form may also present as asymptomatic lymphadenopathy.³

The investigation of choice will be a contrast enhanced CT which usually shows a homogenous mass with uniform enhancement (as in this case- castleman's disease with hyaline vascular type).⁸

An excision biopsy is necessary to clinch the diagnosis as an FNAC will usually be inconclusive. An excision biopsy maintains the germinal centre and the interfollicular zone which is essential for an accurate diagnosis.⁹

Usually the hyaline vascular type of Castleman's disease is completely cured by excision biopsy (comprising most unicentric disease), with no known cases of recurrence documented in the literature. The plasma cell type showed one case of recurrence reported by Sanz et al in a case of Castleman's disease in the head and neck region.¹⁰

The multicentric type after obtaining a diagnosis is treated with chemotherapy, radiation and immunomodulators including steroids, rituximab and anti-IL-6 antibodies.¹¹

When dealing with neck masses, apart from suspecting lymphoma, secondary deposits and tuberculous lymphadenitis one must also maintain a high degree of suspicion for differentials like Castleman's disease. The CT picture will also aid us as Castleman's disease usually has a homogeneous enhancement (hyaline vascular type) but areas of necrotic lymphadenopathy presents with only ring enhancement. In conclusion, Castleman's disease must be considered as a differential diagnosis when dealing with lymphadenopathy in the head and neck region.

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REFERENCES

1. Castleman B, Iverson L, Menerdez VP. Localized mediastinal lymph node hyperplasia resembling thymoma. *Cancer*. 1956;9:822-30.
2. Keller AR, Hochlhalzer L, Castleman B. Hyaline vascular and plasma cell types of giant lymph node hyperplasia of the mediastinum and other locations. *Cancer*. 1972;29:670-83.
3. Jongsma TE, Verburg RJ, Geelhoed-Duijvestijn PH. Castleman's disease: a rare lymphoproliferative disorder. *Eur J Intern Med*. 2007;18:87.
4. Abell MR. Lymphoid hamartoma. *Radiol Clin North Am*. 1968;1:15-24.
5. Lida E, Kohno A, Mikami T, Kumekawa H, Akimoto S, Hamano K. Mesenteric Castleman tumor (case report). *J Comput Assist Tomogr*. 1983;7:338-40.
6. Koslin OK, Borland LL, Sekar BC. Cervical Castleman disease: CT study with angiographic correlation. *Radiology*. 1986;160:213-4.
7. Ferreiros J, Gomez-Leon N, Mata MI, Casanova A, Pedrosa CS, Cuevas A. Computed tomography in abdominal Castleman's disease. *J Comput Assist Tomogr*. 1989;13:433-6.
8. Ko SF, Hsieh MJ, Ng SH, Lin JW, Wan YL, Lee TY, et al. Imaging spectrum of Castleman's disease. *AJR Am J Roentgenol*. 2004;182:769-75.
9. Sudha A, Vivekanand N. Cytologic picture of Castleman's disease: a report of two cases. *J Cytol*. 2010;27:152-4.
10. Sanz C, Sierra J, Cobarro J, Avellaneda R, Montserrat E, Rozman C. An unusual case of Castleman's disease restricted to the neck. *ORL J Otorhinolaryngol Relat Spec*. 1992;54:331-3.
11. Talat N, Schulte KM. Castleman's disease: systematic analysis of 416 patients from the literature. *Oncologist*. 2011;16:1316-24.

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