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Case Report

A rare case report of hyaline vascular variant of Castleman's disease

Sarat Manohar[1], **Naveen Chand**[1], **Monika Mishra**[1], **Gunvanti Rathod**[1]*

[1]*Dept. of Pathology, AIIMS - All India Institute of Medical Sciences, Bibinagar, Telangana, India*

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ABSTRACT

Castleman's disease, is a heterogenous spectrum of lymphoproliferative disorder also known as angiofollicular lymph node hyperplasia or giant lymph node hyperplasia, offers a diagnostic dilemma enriched with histological differences. Its manifestations, range from localized to systemic manifestation with progression to fatal disease involving multiple organs. It has 3 histological types- hyaline vascular variant (HVCD), plasma cell variant (PCCD) and mixed hyaline vascular and plasma cell variant. Clinically it can be classified as unicentric and multicentric. Here we report a case of unicentric Castleman disease in a 55 year old woman who presented with a swelling in the posterior aspect of the neck which was radiologically diagnosed as lipoma and histologically to proven to be a unicentric hyaline vascular type of Castleman's disease.

1. Introduction

Castleman's disease, is a heterogenous spectrum of lymphoproliferative disorder also known as angiofollicular lymph node hyperplasia or giant lymph node hyperplasia, offers a diagnostic dilemma enriched with histological differences.[1,2] Its manifestations, range from localized to systemic manifestation with progression to fatal disease involving multiple organs. It has 3 histological types- hyaline vascular variant (HVCD), plasma cell variant (PCCD) and mixed hyaline vascular and plasma cell variant. Clinically it can be classified as unicentric and multicentric.[1,3] Nearly all cases of unicentric Castleman disease are of hyaline-vascular type; hyaline-vascular Castleman disease has well-defined pathological features, and likely represents a neoplasm of stromal origin with abundant associated reactive lymphoid tissue. Multicentric Castleman disease (MCD) includes cases related to KSHV/HHV8 infection (KSHV/HHV8+ MCD) and cases of uncertain etiology (idiopathic MCD, iMCD). A subset of cases of iMCD fulfills criteria for TAFRO (Thrombocytopenia, Anasarca, Fever/inflammatory symptoms, Renal dysfunction/bone marrow reticulin fibrosis, Organomegaly).

The unicentric form presents commonly as a slow-growing solitary mass occurring usually in the body cavities like mediastinum, abdominal cavity, retroperitoneum, pelvis as well as neck. Most of these lesions are asymptomatic and identified incidentally by the clinician. In such cases, surgery is the treatment of choice and has a curative potential. In contrast, MCD affects multiple lymph nodes all over the body and these patients usually have severe symptoms and are treated by a hematooncologist usually after lymph node harvesting.[4] The aim of this rare case reporting is to describe a spotted case of Unicentric Castleman's disease with all classical histological findings and its diagnostic tools.

* *Corresponding author*.
E-mail address: neempath@gmail com (G. Rathod).

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2. Case Report

A 55 year old woman presented with complaint of a swelling over nape of neck, which was present since 3 years and gradually increasing in size. On Clinical evaluation a soft, mobile, non tender 2x2 cm size swelling over nape of neck was present. No punctum or any discharge from swelling noted. Apart from this lesion the patient also had another swelling over lower back which was diagnosed histologically as epidermal inclusion cyst. The lesion was excised and sent for histopathological evaluation. Grossly a 2.5x2.0x1.5 cm grey white nodule was received which on cut section it was solid and grey white. Histopathological examination showed a lymph node architecture with hyperplastic follicles with many follicles showing prominent mantle zones with onion skin like appearance. There was also presence of twinning of follicles and the follicles were traversed by sclerotic and hyalinised vessels imparting lollipop like appearance. The expanded interfollicular areas show extensive vascular proliferation with hyalinization along with many eosinophils, immunoblasts and few plasma cells.(Figures 1, 2, 3 and 4) It was reported as Hyaline vascular type of Castleman's disease. On further evaluation the patient had no fever, weight loss, anasarca, generalized lymphadenopathy or organomegaly.

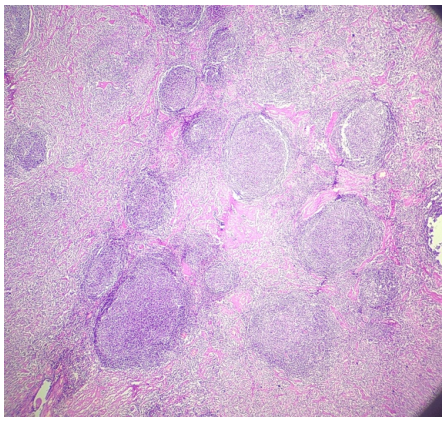

Figure 1: Showed hyperplastic follicles (4 X, H & E stain)

3. Discussion

Castleman disease is a heterogenous group of diseases with variable clinical presentations and prognosis.[5] In 1954, Benjamin Castleman described a 40-year-old man who presented with many years of fever, weakness, and nonproductive cough and was found to have a large mediastinal mass at fluoroscopy. It has 3 histological types- hyaline vascular variant (HVCD), plasma cell variant (PCCD) and mixed hyaline vascular and plasma cell variant. Clinically it can be classified as unicentric and multicentric.[1] It can be unicentric or multicentric

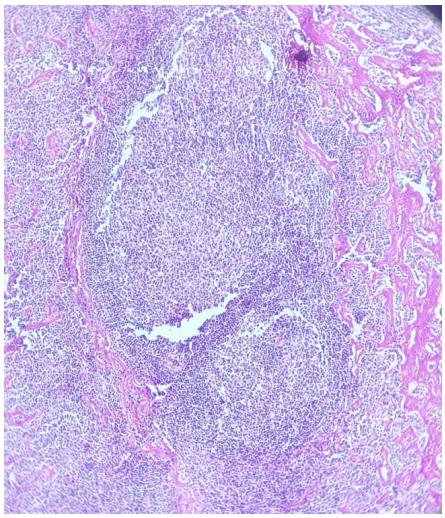

Figure 2: Showed twinning of lymphoid follicles (10 X, H & E stain)

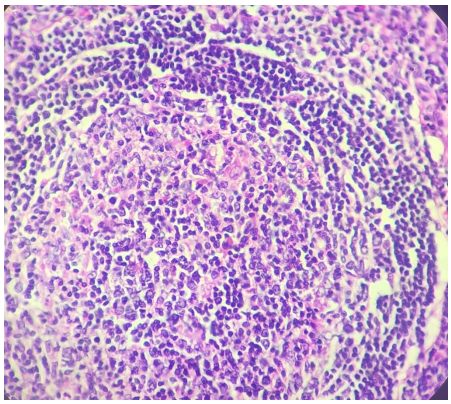

Figure 3: Showed prominent mantle zone with onion skin appearance (40 X, H & E stain)

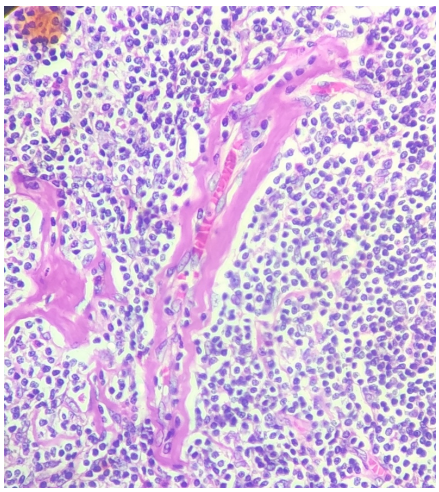

Figure 4: Showed prominent hyalinised vessel (40 X, H & E stain)

and UCD presents commonly as a solitary asymptomatic slow growing mass. The common type, Classic Hyaline-vascular form usually presents as unicentric disease with single lymph node in the age group of 40 to 60 years mostly in thoracic region.[2] In the present case, 55 years old female patient presented with swelling at nape of neck since 1 year. The etiology is unclear except in HHV -8 associated subtypes. Several studies tried to highlight the etiopathogenesis of the disease and some of which have noted an overexpression of IL-6 leading to vasculogenesis.[6–8]

The risk factors for Castleman disease are thymoma, organ transplant, recent vaccination, intrauterine device, consanguineous parentage and viral infection as HHV-8, HIV, HCV.[1,6] Radiological examination with CE-CT shows a marked venous phase enhancement due to rich vascularity and decreased enhancement observed in late phases due to the escape of contrast agent into the extravascular space. Histologically it has to be differentiated from lymph nodal lesions ranging from reactive hyperplasia to lymphoproliferative disorders. The WHO classification of hematolymphoid tumors has delineated major and minor criteria to diagnose these variants. The unicentric hyaline vascular variant requires grade 2-3 regressed follicles/hyaline vascular follicles, prominent stroma, prominent high endothelial vessels and relatively few plasma cells. All the above mentioned features are essential and need to seen to make the diagnosis.[5,9] In the present case we had noticed all these classic microscopic features of hyaline vascular variant of Castleman's disease. Castleman's disease can be associated with other hematologic diseases such as myeloma, lymphoma, amyloidosis or POEMS syndrome. Individual cases have to be cautiously approached keeping in view of the above mentioned associations and correlation with relevant biochemical parameters is essential. Surgical resection is considered the cornerstone of radical treatment for unicentric CD and is the most widely accepted therapy in the literature therefore the upfront excision decision was taken in our case for diagnostic and radical concerns. In case the surgical resection was not applicable, or the lymph node is not accessible, radiation therapy could be an effective way to destroy the affected tissue.

4. Conclusion

This case of Castleman's disease is presented because of its rare incidence and presence of all classical microscopic findings of Hyaline vascular variant. It is a lymphoproliferative disorder with a benign clinical course. The Hyaline vascular variant is commonly seen. Surgical resection of the tumors in the unicentric localized type of CD is the Diagnostic & Therapeutic Procedure of choice. We should never miss the fact that all patients diagnosed with CD should receive a systemic survey to exclude the possibility of ignored lesions or double pathologies and the importance of regular follow ups to monitor the possibility of recurrence.

5. Source of Funding

None.

6. Conflict of Interest

None.

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Author biography

Sarat Manohar, Senior Resident https://orcid.org/0009-0009-2137-3678

Naveen Chand, Senior Resident https://orcid.org/0009-0005-8228-3052

Monika Mishra, Senior Resident

Gunvanti Rathod, Additional Professor https://orcid.org/0000-0002-1045-8707

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