

Castleman Disease with Endocrine Dysfunction

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Article History	Abstract
Received: 06 June 2023 Revised: 05 Sept 2023 Accepted: 25 Nov 2023	Castleman disease is a rare, non-clonal lymphoproliferative disorder predominantly affecting lymphoid tissues. This report presents the case of a 23-year-old male with a chronic left-sided neck swelling and gynecomastia. Surgical excision and Webster technique for gynecomastia were performed, and histopathological examination revealed features consistent with Castleman disease. The lymph node specimen displayed enlarged and reactive atretic germinal centers with an onion skin arrangement of lymphocytes and occasional atypical follicular dendritic cells. Additionally, the breast tissue showed proliferating ducts indicative of gynecomastia. Immunohistochemical analysis confirmed the diagnosis, revealing CD20-positive mantle zones, diffuse CD45 positivity, CD3 positivity in the paracortex and interfollicular area, and Ki-67 positivity in the germinal center. Castleman disease presents clinically as unicentric or multicentric forms and histologically as hyaline vascular, plasma cell, mixed cellular, or plasmablastic variants. Elevated levels of vascular endothelial growth factor (VEGF) and interleukin-6 (IL-6) underscore the disease's inflammatory background. The disease's heterogeneity presents challenges in diagnosis, particularly the plasma cell variant, which can mimic Hodgkin lymphoma. Laboratory abnormalities, such as elevated ESR, LDH, CRP, and anemia, are seen in the multicentric variant. Associations with POEMS syndrome, Kaposi's sarcoma, and co-existing malignant lymphomas complicate the disease's clinical course. Recognition of this condition is crucial for timely intervention, as surgical excision is curative in unicentric cases, while multicentric forms often exhibit a rebound effect. Understanding the disease's diverse presentations and associated lymphomas is essential for accurate diagnosis and appropriate management.
CC License CC-BY-NC-SA 4.0	Keywords: Castleman Disease, Angiofollicular Lymph Node

1. Introduction

Castleman disease is a rare disease of unknown etiology. It is non-clonal lymphoproliferative disorder. It occurs most commonly in adults, but it can also affect children. Castleman disease most commonly affects lymphoid tissues in thorax, abdomen, pelvis and neck.

2. Case Report

A 23 year old male came with complaints of swelling in left side of Neck since childhood, No h/o loss of weight /loss of appetite, No h/o pain. L/E: swelling of size 4x3cm in left side angle of mandible, left side of chest: Gynecomastia present. Surgery: Excision done for swelling and webster technique for gynecomastia sent for HPE. Received two specimens in separate containers, one containing grey white lymph node and other containing multiple yellowish fatty pieces. histopathology and immunohistochemical study had been done. Histopathological examination of one specimen revealed portions of enlarged and reactive lymph node composed of atretic germinal centers with expanded mantle zones. the lymphocytes are arranged in a onion skin pattern (figure 1.3) around the germinal centers. the germinal centers show follicular dendritic cells and occasional plasma cells with atypical follicular dendritic cells at places. The twinning of germinal centers is seen at places (figure 1.1). The vascular channel penetrating the mantle zone giving lollipop appearance is seen occasionally (figure 1.2). Suggestive of castleman's disease. The other specimen section shows portions of fibrofatty tissue showing proliferating ducts Gynecomastia of left breast

A panel of IHC markers used to confirm the diagnosis

CD 20 – Positive in Mantle Zone (Figure 1.4)

CD 45 – Diffuse Positive

CD 3 – Positive in Paracortex and Interfollicular Area

Ki 67 – Positive in Germinal center

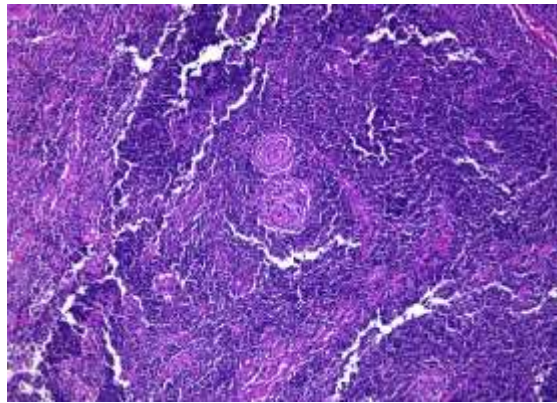


Figure 1.1

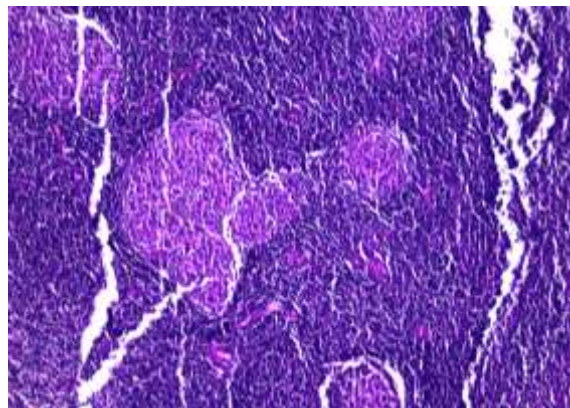


Figure 1.2

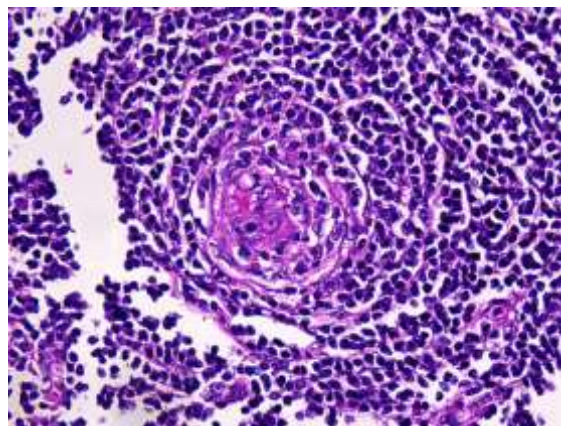


Figure 1.3

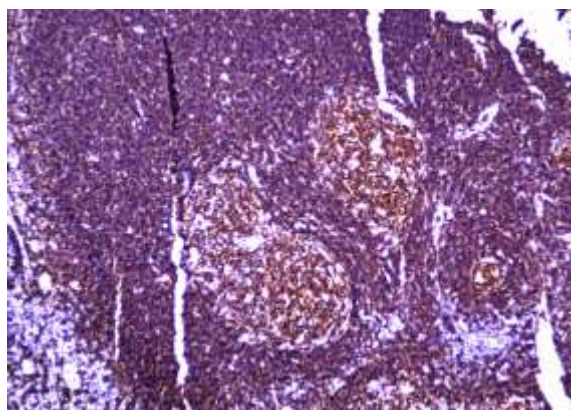


Figure 1.4

3. Results and Discussion

Castleman disease can be classified clinically into a unicentric or multicentric form, depending on the number of lymph nodes involved, and histologically into a hyaline vascular variant, plasma cell, mixed cellular, or plasmablastic variant. The disease has a predominantly inflammatory background, reflected in high levels of vascular endothelial growth factor (vegf) and interleukin-6 (il-6). The role of cytokines in this disease explains the clinical presentation. The clinical scenario varies widely, based mainly on the histologic type. Plasma cell variant- castleman disease obscure the neoplastic cells of Hodgkin lymphoma and delay correct diagnosis. There is elevated laboratory abnormalities such as Anemia, Elevated ESR, LDH, CRP in multicentric variant. Some associated diseases are POEMS syndrome and kaposi's sarcoma. Malignant lymphomas can also co-exist in patients with castleman disease. In HHV-8- patients, classical hodgkin's lymphoma, diffuse large B-cell lymphoma, mantle cell lymphoma are most often reported. In HHV-8+ patients, primary effusion lymphoma and plasmablastic lymphoma are commonly reported.

4. Conclusion

This is one of the reasons that the incidence and prevalence of the disease are not only underestimated but has not even been well-established. Therefore, it is important that physicians consider this clinical entity so patients can receive adequate and early treatment. Treatment usually depends on the type of disease; in unicentric has good response on surgery but multicentric has rebound effect.

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Conflicts of interest

There are no conflicts of interest

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