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Castleman's Disease - A rare case presentation as cervical lymhadenopathy

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Abstract

Introduction: Castleman's Disease (CD) is a rare, non clonal lymphoproliferative disorder having distinct subtypes depending on its aetiology, pathology, and clinical presentation. Pathologically it can be classified as Hyaline Vascular Type (HV-CD), plasma cell type, mixed type, and Human Herpes virus (HHV)-8 associated Castleman's disease. Clinically it manifests as the more common unicentric (localized or unifocal) Castleman's disease (UCD) and the less common multicentric (generalized or multifocal) Castleman's disease (MCD)[1].

Case Report: A 16 year old female presented with complaints of swelling in the left side of the neck since 5 years that gradually increased in size and was painless with no history of loss of weight or evening rise of temperature.

Diagnosis: Unicentric Castleman's Disease.

Conclusion: This case report is presented for its rarity. Neck lymph nodes are involved by CD and may be confused with other common causes of neck lymphadenopathy like tuberculosis and nodal secondaries. Surgical removal of the tumors in the unicentric type of CD is the treatment of choice.

Keywords: Cattleman's Disease, Unicentric Cattleman's Disease, Multicentric Cattleman's Disease, Human Herpes Virus, Hyaline vascular type

Introduction

In 1956, Castleman and his colleagues first described the clinicopathologic entity of mediastinal lymphoid hyperplasia with hyalinization of follicles and interfollicular vascular formation. Castleman's Disease (CD) is an uncommon disorder that can involve either a single lymph node (unicentric- UCD) or multiple lymph nodes (multicentric- MCD) [1].

The estimated CD incidence is approximately 25 cases per million person-years, which represents under 5200 cases in the United States per year[2].

A 2008 study by Simpson reported that:

- Unicentric Castleman's Disease (UCD) occurs at a rate of 16 per million patient-years and can affect individuals of all age groups.
- The incidence of HHV-8-associated CastlCast's Disease (CD) varies significantly, with a higher prevalence among HIV-positive men.
- Idiopathic Multicentric Castleman Disease (iMCD) has an incidence of 5 per million patient-years.[3]

Additionally, a US-based study (2000–2009) analyzing 59 MCD patients found that 61% were male, with a mean age of 53 years, and 68% were white[4]

This rare lymphoproliferative disorder has been linked to the Human Herpes virus-8 (HHV 8) that infects both B-cells and the lymphovascular compartment of lymph nodes; however, the etiology and pathophysiology of CD remains elusive and may be related to dysregulation of the immune system[3]. However, IL-6 is associated with UCD, while MCD is associated with HHV infection[4].

Castleman's Disease (CD) describes a group of at least four disorders that share a spectrum of characteristic histopathological features but have a wide range of aetiologies, presentations, treatments, and outcomes[5].

CD includes unicentric CD (UCD) and multicentric CD (MCD), the latter of which is divided into idiopathic MCD (iMCD), Human Herpes Virus-8 (HHV8)-associated MCD (HHV8-MCD), and polyneuropathy, organomegaly, endocrinopathy, monoclonal plasma cell disorder, skin changes (POEMS)-associated MCD (POEMS-MCD). iMCD can be further subclassified into iMCD–thrombocytopenia, ascites, reticulin fibrosis, renal dysfunction, organomegaly (iMCD-TAFRO) or iMCD–not otherwise specified (iMCD-NOS). The advent of effective retroviral therapy and use of rituximab in HHV8-MCD have improved outcomes in HHV8-MCD. Anti–interleukin-6–directed therapies are highly effective in many iMCD patients, but additional therapies are required for refractory cases.[6]

Recently, the Castleman's Disease Collaborative Network (CDCN) proposed a classification system retaining the UCD vs MCD nomenclature but further dividing MCD by aetiological driver (HHV8-associated MCD [HHV8-MCD]; POEMS-associated MCD [POEMS-MCD]; iMCD) and within iMCD by phenotype, iMCD-TAFRO, and iMCD–not otherwise specified (iMCD-NOS)[6].

Case study

Case: A 16 year old young female patient came to the General surgery Outpatient Department with complaints of swelling in left side of neck since 5 years that was noticed by patient's father which was insidious in onset, initially the size of a peanut, which gradually increased to the present size of 2x3 cm and was not associated with any pain. No history of loss of weight or evening rise of temperature. No significant past history or tuberculosis contact.

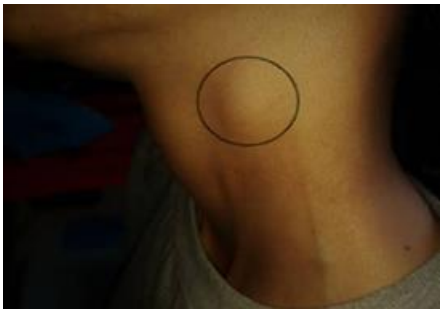

Figure 1: Preoperative swelling noted over the left side of the neck

No history of similar swelling complaints in the past.

Clinical examination

Inspection: Solitary swelling noted over the left side of neck over the anterior border of left sternocleidomastoid. All margins well defined, had a smooth surface, extending from around 3cm above clavicle to 5 cm cranially and horizontally from anterior border of sternocleidomastoid to 2cm laterally.

Palpation: multiple discrete swellings situated over left side of neck largest measuring around 3x2 cm, smooth surface, with well-defined border, extending from around 3cm above clavicle to 5 cm cranially and horizontally from anterior border of sternocleidomastoid to 2cm laterally, soft to firm in consistency, non tender, mobile ,with no skin changes. No visible pulsations. Level 2 and level 3 lymph node groups were enlarged.

On CECT: A well-defined enhancing lesion measuring 50x35x26mm noted in the left side of neck beneath the left sternocleidomastoid muscle with no muscle infiltration. Lesion noted at C2-C4 vertebra level with multiple enlarged left level II and III lymph nodes which shows similar enhancement to lesion.

She underwent excision of lymph node with swelling under General Anesthesia.

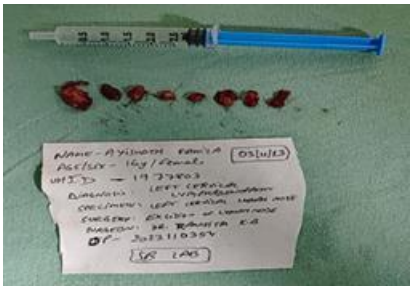

Figure 2: postoperative specimen with multiple enlarged lymphnodes (Level 2 and 3)

On HPE: Benign reactive lymphnode with follicular hyperplasia; multiple fragmented lymphoid tissue showing reactive lymphoid follicle with numerous macrophages and many polymorphs. Inter follicular space shows lymphoid cells and few plasma cells, along with sinus histiocytes and congested vascular channels. There is no evidence of granuloma or malignancy.

Other routine investigations were found to be within normal limits.

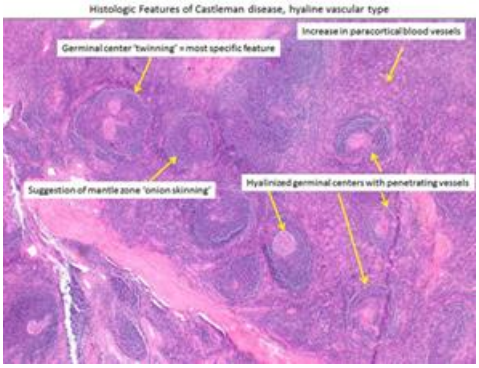

Figure 3: HPE picture depicting Castleman's onion skin like or lollipop sign.

Immuno-Histo-Chemistry (IHC): IHC positive for CD20, CD3, CD23, MUM1, PAX5.

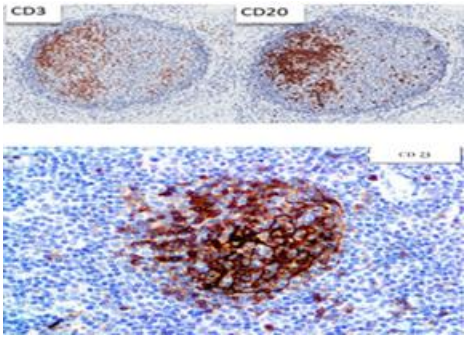

Figure 3: IHC marker study positive for CD3, CD20 CD23

Discussion

CD can present as either a unicentric (one site involvement) or a multicentric disease (more than one site involvement).[1]

On the basis of histopathology, four subtypes are found, namely, the hyaline-vascular variant, plasma cell variant, mixed-cell type, and plasmablastic type (plasmablasts expressing Human Herpes Virus-8).[2]

The hyaline vascular (HV) type of Castleman's disease is characterized by an increased number of lymphoid follicles with a broad mantle zone and a hypoplastic germinal center. The centrally located germinal center is encircled by small lymphocytes arranged in concentric layers, forming an "onion skin-like" pattern[5]. Hyalinized capillaries penetrate the center of these follicles, creating the distinctive "lollipop sign."[6] The interfollicular zones exhibit prominent endothelial vascular proliferation and hyalinization, along with variable numbers of small lymphocytes, immature plasma cells, and dyspattern Follicular Dendritic Cells (FDCs). A high concentration of plasmacytoid dendritic cells is often found around these lesions[7].

In plasma cell Castleman's Disease (PC-CD), mature lymphocyte infiltration is present, while the mantle zone and germinal center proportions remain intact. The interfollicular regions are densely populated with mature plasma cells and are less vascular than in HV-CD. Occasionally, irregular clusters of polyclonal plasmablasts may be observed within the mantle zone of B-lymphocytes[8].

The mixed type exhibits feature of both HV-CD and PC-CD.HHV-8-associated Castleman's Disease is characterized by intense angiogenesis between lymphoid follicles with indistinct boundaries and the presence of immature plasmablasts, which may exhibit restricted lambda light chain expression in specific immunoglobulins[9].

Unicentric Castleman's disease can be cured by surgically removing the diseased lymph node[9].

Multicentric Castleman disease may include:

Immunotherapy: The use of drugs such as siltuximab (Sylvant) or rituximab (Rituxan) can block the action of a protein that is made in excess in people who have multicentric Castleman's Disease[9].

Corticosteroids: Drugs such as prednisone can help control inflammation[10].

Antiviral drugs: These drugs can block the activity of HHV-8 or HIV if the patient has one or both of those viruses[10].

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