Case Report

Castleman's Disease of the Chest Wall Masquerading as a Breast Mass

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Abstract

Castleman's disease (CD) is a rare lymphoproliferative disorder with unknown etiology and is clinically associated with abnormal lymph node enlargement. CD is classified clinically as unicentric CD (UCD) or multicentric CD (MCD) and histologically as hyaline vascular variant (HVV) or plasma cell variant (PCV). Primary axillary localization of CD represents 2% of the cases. CD rarely occurs in the breasts. The CD is easily confused with lymphoma or other solid tumors. However, the infrequent occurrence of CD has led to a limited analysis of its clinical presentation and susceptibility to misdiagnosis during preliminary investigations. Therefore, accurate diagnosis requires clinical, radiological, and histopathological evaluation. We herein describe a rare case of CD in a 38-year-old female who initially presented with a swelling in the right breast. CECT as well as FNAC of the lesion was suggestive of a mesenchymal tumor. The patient underwent axillary-wide local excision.

Keywords: Castleman Disease; Lymphoproliferative Disorders; Unicentric Castleman Disease; Hyaline-Vascular Type.

Introduction

Castleman disease (CD), also known as angio-follicular or benign giant lymph node hyperplasia, is a complex lymphoproliferative disorder broadly divided into two forms: unicentric CD (UCD) and multicentric CD (MCD) [1]. The former is typical of the hyaline-vascular type and amenable to surgical treatment, while the latter normally belongs to the plasma cell type and is associated with more complicated systemic manifestations [2]. The CD was first described by Benjamin Castleman in 1954 [3]. The cause of CD remains unclear; however, possible causes include chronic inflammation, lymphoid hamartomatous hyperplasia, and an increase in serum interleukin-6 levels, among others [4]. CD most commonly affects lymph nodes, arising in nodal stations throughout the body but most commonly involving the chest region [5]. MCD often presents with generalized lymphadenopathy and systemic symptoms such as fever, night sweats, weight loss, and fatigue, closely resembling the clinical picture of malignant lymphomas [6]. UCD typically manifests as a solitary, asymptomatic mass, frequently located in the mediastinum. Due to its location and imaging characteristics, it can be mistaken for other mediastinal tumors, including neurogenic tumors [7]. Furthermore, each subtype of CD has varying clinical features, etiologies, treatments, and outcomes. Hence, an accurate diagnosis requires a combination of clinical, radiological, and histopathological findings.

Castleman disease (CD) exhibits a range of histological patterns [8]. The hyaline vascular type is characterized by atypical follicles with reduced germinal centers, surrounded by broadened mantle zones of small lymphocytes arranged in concentric "onion-skin" layers [8]. These germinal centers may show hyalinization and contain follicular dendritic cells [8]. In contrast, the plasma cell type presents with enlarged germinal centers and a notable accumulation of polyclonal plasma cells in the interfollicular spaces [8]. Both types display prominent vascular proliferation in the interfollicular areas [8]. The mixed variant shows features of both hyaline vascular and plasma cell types. A less common form, the plasmablastic variant, is associated with HIV infection. This subtype, a variant of the plasma cell form, includes large plasmablasts infected with human herpesvirus 8 (HHV-8) and has the potential to progress into a monoclonal plasmablastic lymphoma [8]. Diagnostically, these histological features can be challenging, as they are not exclusive to Castleman disease and may also appear in other conditions.

Case Report

A 38-year-old female presented to the outpatient clinic at PGIMS, Rohtak with a chief complaint of swelling in her right breast. On examination, the swelling was nontender, gradually progressive, and associated with discomfort and generalized fatigue over the past 8 months. Past Medical History, No significant medical history. Physical Examination: Single, enlarged, non-tender swelling in the upper outer quadrant of the right breast measuring 5 $\times$ 4 cm.

Investigations: Ultrasound of the right breast revealed a well-defined oval heterogeneously hypoechoic lesion measuring 25 $\times$ 23 mm in the right anterior chest wall region at 11 to 1 o'clock position (Fig. 1). The lesion showed significant vascularity on color Doppler. CECT chest revealed a well-circumscribed oval-shaped altered signal intensity lesion measuring approximately AP 23 $\times$ TR 53 $\times$ CC 75 mm in the right upper chest wall in the intermuscular plane of pectoralis major and minor with no obvious infiltration of muscle fibers and no obvious marrow edema or destruction of underlying ribs; suggestive of mesenchymal tumor (Fig. 2). USG-guided FNAC of the lesion yielded blood mixed aspirate and the stained smears examined were mildly cellular and showed the presence of isolated and few clusters of round to oval spindle cells along with lymphoid cells in the hemorrhagic background. These features were reported as suggestive of mesenchymal neoplasm and biopsy and IHC was advised for confirmation and further typing.

Laboratory Tests: All laboratory investigations including CBC, LFT, KFT, serum electrolytes, ECG, and lipid profile were within normal limits. Viral markers were non-reactive.

Operative Procedure: Axillary-wide local excision was done. Lazy-S incision was made in the right axilla. A well-encapsulated 10 $\times$ 5 cm lesion was found in the interpectoral plane. The rest of the visualized structures were normal. The mass was resected out and vessels supplying were ligated and cut. Subcutaneous tissue closed with Vicryl 3-0 RB. Skin closed with a stapler. Hemostasis was achieved at each step. Antiseptic dressing (ASD) was done (Fig. 3).

Histopathological examination and Immunohistochemistry (IHC): Grossly, we received an encapsulated bean-shaped soft tissue piece measuring 7 $\times$ 5 $\times$ 3 cm. The external surface was unremarkable. On the cut section, homogenous grey-white areas were identified (Fig. 4). On microscopy, sections examined showed the structure of a lymph node with characteristic "onion-skin" layering of mantle zone lymphocytes, atrophic germinal centers within the follicles, traversed by blood vessels, and hyalinization (Fig. 5). On IHC, CD3 was positive in the inter-follicular area, BCL2 positivity was seen in mantle zone while it was negative in germinal centre, CD20 was seen positive in follicular cells while CD21 positivity was recorded in follicular dendritic cells (Fig. 6 A, B, C and D respectively). Diagnosis: Unicentric Castleman disease; hyaline vascular type.

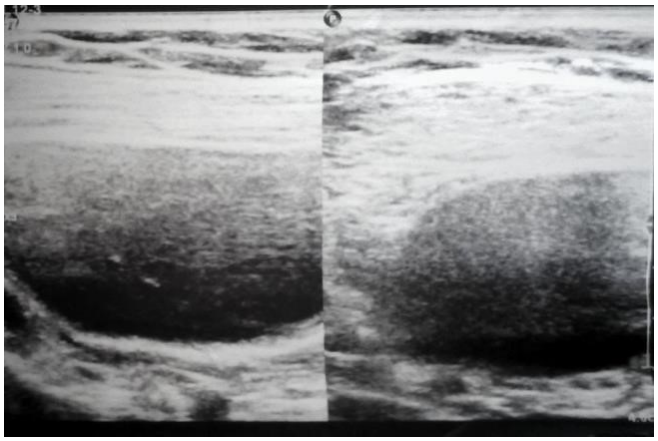

Figure 1: USG findings. A well-defined oval heterogeneously hypoechoic lesion measuring 25 $\times$ 23 mm in the right anterior chest wall region at 11 to 1 o'clock position. The lesion showed significant vascularity on color Doppler.

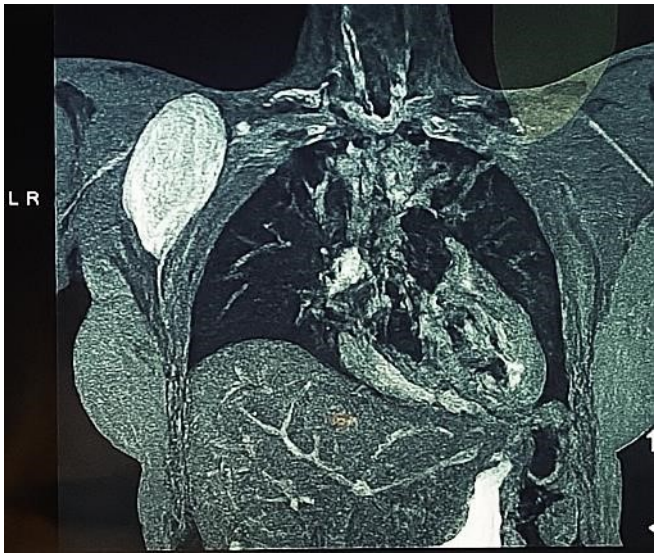

Figure 2: CECT chest. A well-circumscribed oval-shaped altered signal intensity lesion in the right upper chest wall in the intermuscular plane of pectoralis major and minor.

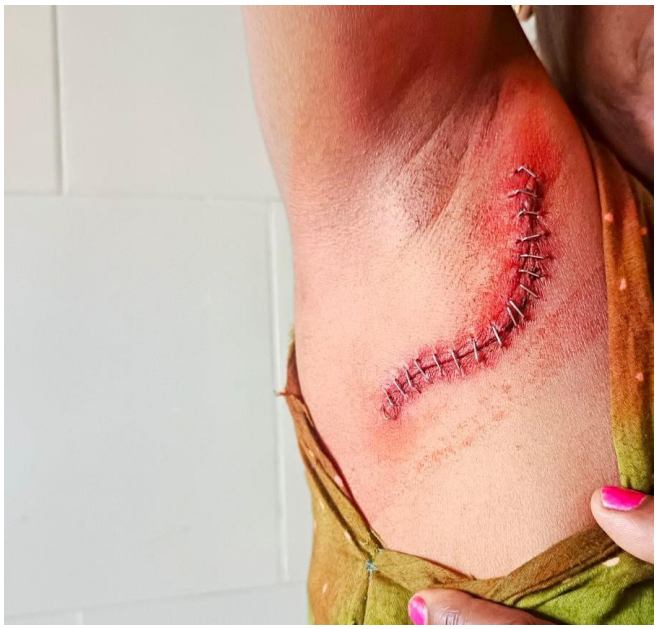

Figure 3: Post-operative site of excision.

Discussion

Castleman Disease (CD) is characterized by abnormal lymphoid tissue growth, with distinct presentations in the form of Unicentric (UCD) and Multicentric (MCD) classifications. Typically, 90% of unicentric CD cases are either asymptomatic or present with compressive symptoms due to mass effect, with type B symptoms (fever, night sweats, weight loss) and laboratory anomalies being uncommon [9]. Conversely, multicentric Castleman disease is associated with systemic symptoms. The estimated incidence of UCD is approximately 16 cases per million person-years, with a median age at diagnosis ranging from 30 to 34 years and an equal distribution between genders [9]. UCD is mainly categorized into 2 histological subtypes: the hyaline vascular variant (HV), also referred to as the hypervascular variant and the plasma cell subtype (PV) [8, 9]. The hypervascular variant, which is more prevalent, typically presents with localized lymphadenopathy and is predominantly associated with unicentric Castleman disease [9]. In contrast, the plasma cell subtype, although less common, is characterized by systemic symptoms and multiorgan involvement [8, 9].

The diagnosis of Castleman disease necessitates a comprehensive assessment integrating clinical, radiological, and pathological findings. Treatment strategies, including surgery, radiation therapy, and chemotherapy, are tailored based on the subtype, stage, and extent of the disease [9]. Unicentric Castleman disease (UCD), particularly the hyaline vascular variant is associated with a favorable clinical course. Complete surgical excision often leads to excellent long-term survival rates,

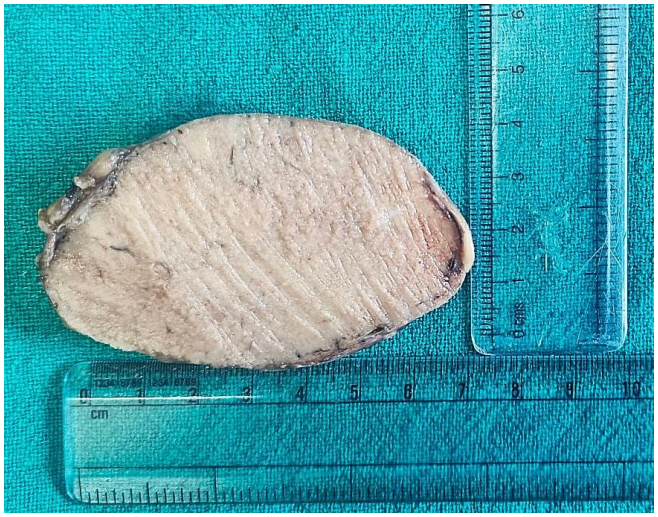

Figure 4: Gross specimen: An encapsulated bean-shaped soft tissue piece measuring 7 × 5 × 3 cm. On the cut section, homogenous grey-white areas were identified.

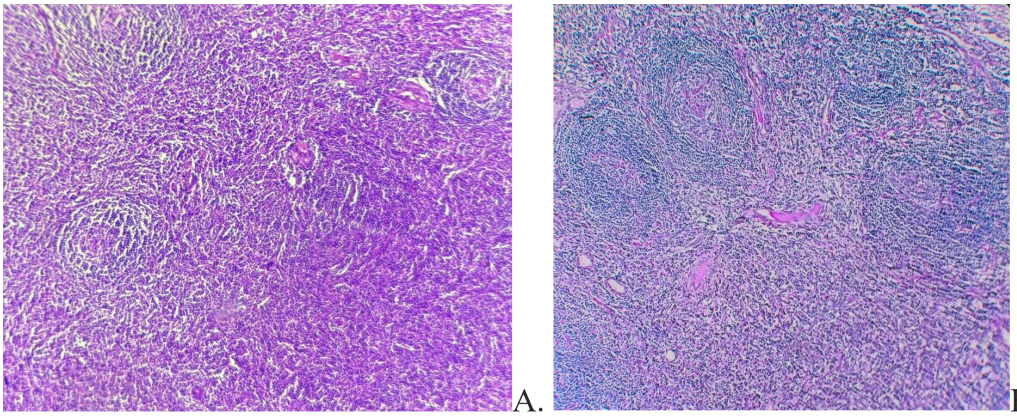

Figure 5: Microscopic Examination (A and B): Characteristic "onion-skin" layering of mantle zone lymphocytes, atrophic germinal centers within the follicles, traversed by blood vessels, and hyalinization. (100X: H&E)

with studies reporting 10-year overall survival rates exceeding 95% [10].

In the current case, the patient presented with a breast lump, the disease is localized to a single lymph node in the right interpectoral region, deviating from the typical presentation of unicentric Castleman disease. On FNAC, a Castleman disease lymph node may appear vascular, spindle-cell rich, and non-lymphoid, especially in the hyaline vascular type, leading to a misdiagnosis of mesenchymal tumor, thereby necessitating tissue biopsy for definitive diagnosis [11]. The location, clinical and laboratory findings of the lesion raised initial suspicions of mesenchymal disease/breast pathology. The final diagnosis of Castleman disease was established from histopathological analysis. Radiological assessments further confirmed the absence of additional lymphadenopathy or involvement of other lymph node regions, confirming the diagnosis of Unicentric CD; hyaline vascular type.

Conclusion

There is a range of clinical presentations of Castleman Disease (CD), and proper diagnosis and treatment depend on knowing the difference between Unicentric (UCD) and Multicentric (MCD) types. In contrast to the conventional mediastinal location, this case highlights the atypical anatomical presentation of unicentric Castleman disease in a female patient which was localized to the right interpectoral region without any systemic symptoms. The patient's subsequent recovery, including symptomatic relief after the intricate surgical excision of the tumor underscore the necessity of a multidisciplinary approach in the management of such rare clinical entities. Further studies are warranted to elucidate the diverse clinical presentations and optimal therapeutic strategies for Castleman disease, especially in uncommon anatomical locations.

Ultimately, this case contributes to the expanding knowledge base in Castleman disease management and lays emphasis on the significance of individualized treatment approaches tailored to the unique characteristics of each patient's presentation.

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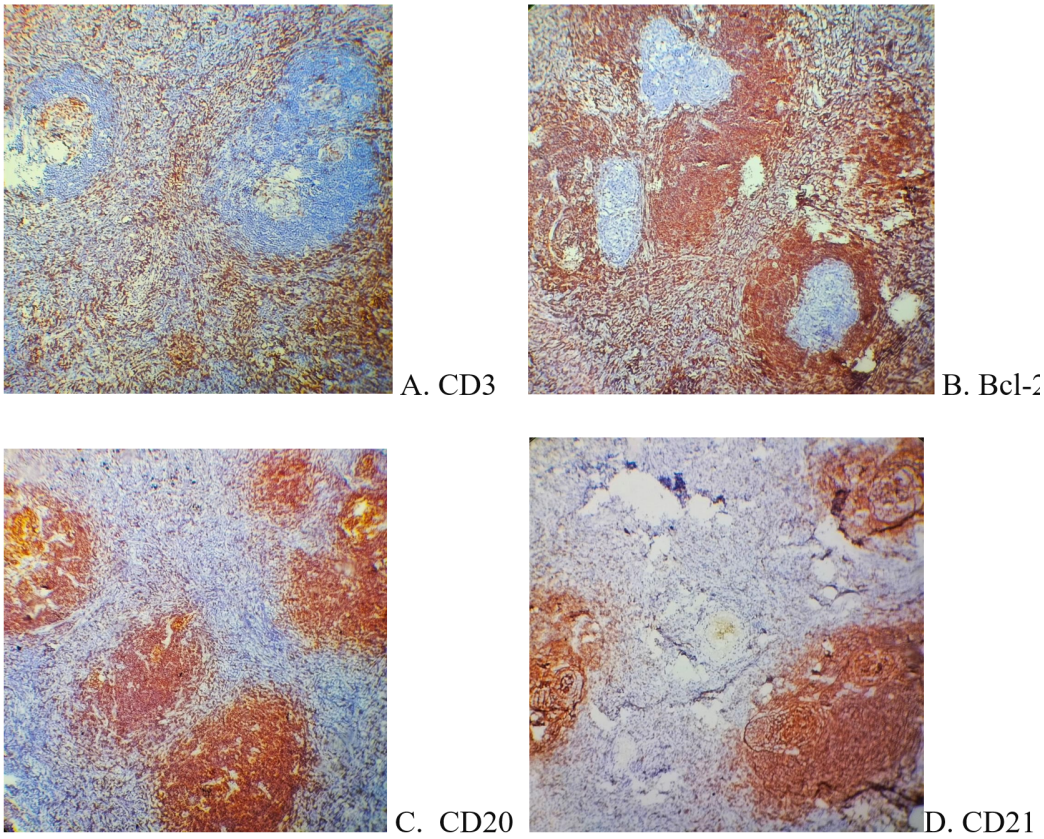

Figure 6: Immunohistochemistry (A,B,C and D): A: CD3 positivity in the inter-follicular area. B: Bcl-2 positivity in the mantle B lymphocytes. C: CD20 positivity in follicular cells. D: CD21 positivity was recorded in follicular dendritic cells (100X; IHC)

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Conflicts of Interest: None.

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