CASE REPORT

A case of Castlemans disease presenting as left axillary tumor

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Introduction
Castleman's disease is an uncommon disease affecting the lymph nodes [1]. It can manifest with a wide range of symptoms that affect people of all age groups. One of the major risk factors for this disease is immunosuppressive states like retrovirus, immunosuppression therapy, etc. We present a case of localised Castleman's disease presenting as left axillary swelling.

Case Report
A 53 year old male presented with a swelling in the left axilla of 1-year duration, which was gradually increasing in size. It was not associated with pain and discharge. On examination, there was a single, non-tender, firm, large mass in the left axilla approximately 8 x 6 cm size, extending behind the pectoralis major muscle (Figure 1). Ultrasound of the left axilla was performed which showed a well-defined hyperechoic lesion measuring 6.2 x 4.0 cm which was suspected to be a fibro-lipoma. To confirm the diagnosis, image guided biopsy was taken from the left axillary swelling which showed occasional reactive follicles with angiofollicular change, parafollicular vascular proliferation, hyalinization with sheets of plasma cells and negative for a focal lesion. We proceeded with MRI scan of the left axilla to find out the extension of the swelling which revealed an enhancing lesion measuring approximately 6.5 x 5.2 cm in the left axillary region, which was found to be abutting the intercostal muscles and ribs, lifting the pectoralis minor anteriorly. The lesion was also displacing the brachial plexus roots, subclavian vessels, with no evidence of chest wall invasion (Figure 2, 3). A differential diagnosis of Neurogenic tumor, was suspected from the findings.

Under general anesthesia, left axillary tumor, was removed successfully (Figure 4). Intraoperatively, it was evident that the tumor was abutting but not involving the brachial plexus or the subclavian vessels and also not invading the muscles. With meticulous dissection, complete removal of the tumor achieved.

Histopathological examination showed lymph nodal tissue with several follicles at the centre of which venules, with
small amount of hyaline were seen with lollipop appearance. There was vascular proliferation and area of fibrosis intersecting the follicles, showing reactive sinus histiocytosis. It was diagnosed as hyaline vascular type of Castleman's disease. Later on, he was evaluated and tested for retrovirus which was found to be negative and interleukin-6 levels which were unremarkable. Immunotyping was performed which did not detect monoclonal gammopathy. And the lymphoma panel tests were positive for CD 3, CD 20, CD 10, BCL 6, CD 138, CD 21, CD 23, Kappa, Lambda. Ki-67 was in a lower level of range. BCL 2 was negative. These results were consistent with Castleman's disease. As this could be a systemic disease, PET CT whole body was taken which showed hypodense nodules in both lobes of the thyroid (SUV max 5.6) in right lobe, largest measuring 0.8 x 0.7 cm. Other than thyroid nodule, no other metabolically active disease noted in the body. FNAC of right lobe of thyroid nodule was done and it was reported as atypical cytology (BETHESDA CATEGORY V). He was recommended surgery in view of the atypical cytology for which the patient chose to defer.

Discussion
In 1954, Dr. Benjamin Castleman first described 2 cases of localised mediastinal lymph node enlargement, which showed vascular proliferation with endothelial hyperplasia, lymphoid follicles with germinal center involution. The disease was named after him. It is a rare lymphoproliferative disorder, which can affect any organs in the body. This disease was expected to involve mediastinal lymph nodes. It also involves the axilla, abdomen, pelvis, breast etc. It initiates an inflammatory process, by releasing cytokines leading to organ dysfunction. It is classified as hyaline vascular (75-78%) and plasma cell (22-25%) based on microscopic examination. It can be unicentric or multicentric. Unicentric type will manifest as localised symptoms with regional lymph nodal involvement. It has an excellent prognosis with excision of involved lymph node. Multicentric is an aggressive type often progressing to lymphoma. It presents with generalised systemic symptoms, abnormal blood investigations. It can be idiopathic or associated with Human Herpesvirus (HHV-8) requiring systemic therapy. Recently, multicentric type is further subdivided into HHV-8-associated which is also known as Kaposi sarcoma-associated herpes virus (KSHV) [2], POEMS associated and idiopathic. TAFRO (thrombocytopenia, anasarca, fever, renal dysfunction / reticulin fibrosis, and organomegaly) is a variant of idiopathic multicentric type which exhibits hypogammaglobulinemia [3]. Multicentric forms an inter relationship with HIV, POEMS Syndrome (Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal gammopathy and Skin changes - hyperpigmentation) and interleukin-6 levels. POEMS syndrome is also known as Osteosclerotic myeloma or Takatsuki or Crow-Fukase syndrome. HHV-8 influences the level of production of interleukin-6. The lymph nodes of Castleman's disease exclusively secretes an excess of interleukin-6 from the germinal centre by B cells. There will be abrupt decrease in the interleukin-6 level with the tumor being removed. If the level does not lower, anti-IL-6 antibody like Tocilizumab, Rituximab are used for multicentric type [4].

In our patient, with the evidence of histological examination, we finally confirmed that axillary mass was due to the Castleman's disease. After the procedure, he was evaluated thoroughly without evidence of systemic involvement. Since it was a localized disease, he did not require any further treatment.
Bo-Kyoung Seo et al. reported a similar case of axillary swelling, incidentally detected in a 45-year-old female without any symptoms [5]. Upon further evaluation with CT scan and USG guided biopsy, they had come to a diagnosis of Castleman's tumor. The tumor was excised. According to this study, only 2% of Castleman's disease manifesting with axillary presentation. USG Doppler is essential in differentiating malignant from benign lymph nodes.

Conclusion
Castleman's disease is an atypical disease which can only be confirmed with histological examination. It should be further evaluated with PET CT scan, as it can produce lesions in the lymph nodes anywhere in the whole body and to get the knowledge of its aggressiveness which really helps in deciding the management protocol.

References

Learning Points:
• Castleman's disease is a rare benign lymphoproliferative disorder that can affect any system of the body.
• It is classified as unicentric and multicentric variety. Diagnosis is mainly confirmed by histopathological examination.
• Treatment varies according to the type of the disease. Hence PET CT scan of whole body is needed to differentiate these types.
• Unicentric type requires local treatment whereas multicentric type requires systemic therapy.