CASTLEMAN DISEASE- A RARE CASE

Pawan Kumar Rathod[1], Lokesh Reddy Chigicherla[2], Venkat Ram Reddy Kuchukulla[3], (Brig) Satyanarayana Moorthy R[4], Ramakrishna Reddy Gaddam[5]

[1]Postgraduate Student, Department of Radiology, SVS Medical College and Hospital, Yenugonda, Mahabubnagar.
[2]Postgraduate Student, Department of Radiology, SVS Medical College and Hospital, Yenugonda, Mahabubnagar.
[3]Professor and HOD, Department of Radiology, SVS Medical College and Hospital, Yenugonda, Mahabubnagar.
[4]Professor, Department of Radiology, SVS Medical College and Hospital, Yenugonda, Mahabubnagar.
[5]Professor, Department of Radiology, SVS Medical College and Hospital, Yenugonda, Mahabubnagar.

HOW TO CITE THIS ARTICLE: Rathod PK, Chigicherla LR, Kuchukulla VRR, et al. Castleman disease - A rare case. J. Evid. Based Med. Healthc. 2017; 4(40), 2447-2449. DOI: 10.18410/jebmh/2017/483

PRESENTATION OF CASE

A 55-year-old female patient presented in the month of September 2016 with a painless mass in left submandibular region, which appeared two years ago and progressively increased in size. There were no constitutional symptoms such as fever, night sweating, fatigue and weight loss. On USG, single well-defined oval-shaped homogeneously hypoechoic mass lesion noted in left submandibular region measuring 6.2 x 3.8 cm. On colour Doppler examination showed peripheral parenchymal vascular flow and increased diastolic flow within center of the lesion. Plain and contrast CT examination was done showed single well-defined hypodense oval-shaped mass lesion in left submandibular region displacing vascular structures medially and posteriorly on post-contrast images showed intense enhancement of mass lesion. Biopsy from the lesion revealed structure of lymph node with partial effacement of lymph nodal architecture. Few lymphoid follicles were seen at periphery. The distorted lymph node showed hyalinised blood vessels in centre of follicle surrounded by mantle of mature lymphocytes. Features suggestive of hyaline vascular variant of angiofollicular lymph node hyperplasia also known as Castleman disease.

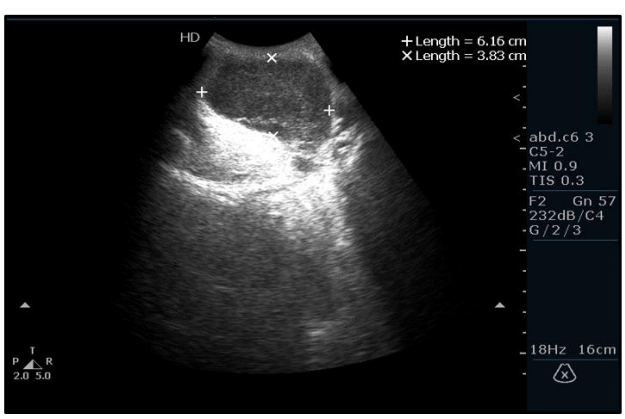

Image 1

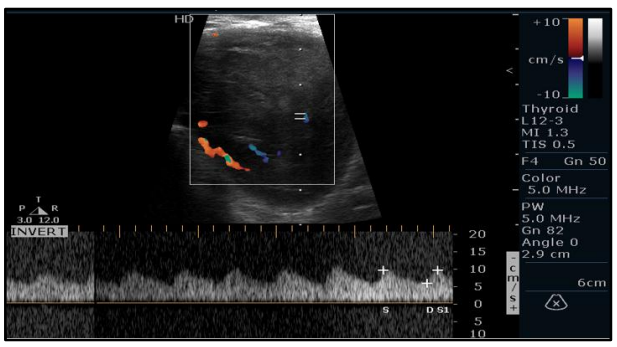

Image 2

- Ultrasonography of neck- Single, well-defined, oval-shaped, homogenous, hypoechoic mass lesion in left submandibular region measuring 6.2 x 3.8 cm in which hilum was not identifiable.
- On colour Doppler examination, lesion showed peripheral parenchymal vascular flow with increased diastolic flow within the lesion.

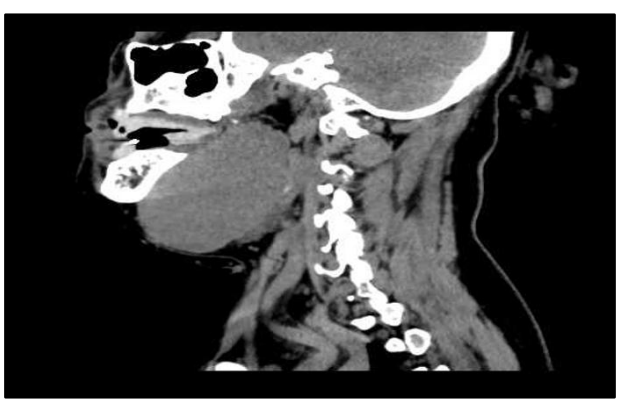

Image 3

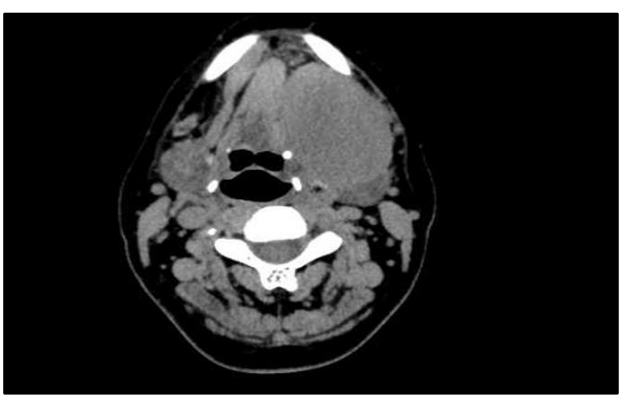

Image 4

Financial or Other, Competing Interest: None.
Submission 28-04-2017, Peer Review 05-05-2017,
Acceptance 16-05-2017, Published 18-05-2017.
Corresponding Author:
Dr. Pavan Kumar Rathod,
C/o. SVS Medical College,
Yenugonda, Mahabubnagar-509001.
E-mail: pavanrathod69@gmail.com
DOI: 10.18410/jebmh/2017/483

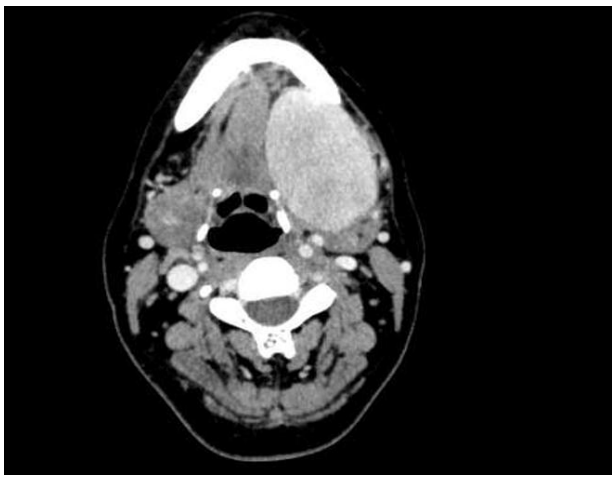

Image 5

CT revealed single, well-defined, oval-shaped, homogenous hypodense mass lesion in left submandibular region measuring 5.7 x 3.5 cm in axial plane, 7.5 x 4.5 cm in sagittal plane and 6.3 x 4.3 cm in coronal plane. Displacing vascular structures medially and posteriorly with no mass effect on larynx. No evidence of bony erosions or destruction.

On post-contrast, images showed intense enhancement.

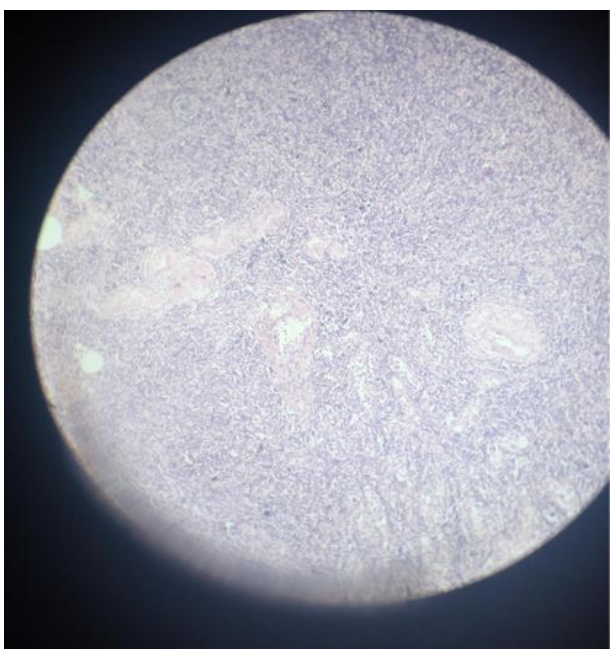

Image 6

Biopsy Findings- From the lesion on the left side revealed structure of lymph node with partial effacement of lymph nodal architecture. Few lymphoid follicles were seen at periphery. The poorly-formed lymph node shows hyalinised blood vessels in centre of follicle surrounded by mantle of mature lymphocytes. Features suggestive of hyaline vascular variant of angiofollicular lymph node hyperplasia also known as Castleman disease.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for painless mass in left submandibular region are reactive lymph nodes, non-Hodgkin's lymphoma, differentiated thyroid carcinoma, carotid body paraganglioma, tuberculosis, sarcoidosis, toxoplasmosis, cytomegalovirus, mononucleosis, HIV or some tumours such as neurofibroma and lymph node metastasis.

PATHOLOGICAL DISCUSSION

The histopathological features of Castleman disease like increase in plasma cells and immunoblasts, germinal centre hyperplasia and increased vascularity are seen as exaggerations of responses to normal antigenic stimuli.

Studies in the early 1990's have found a correlation between the systemic manifestations of unicentric Castleman disease and local production of interleukin-6.[1,2] The exact cells, which seem to produce IL-6 has not been elucidated yet, but candidate cells include follicular dendritic cells, germinal centre B cells or the interfollicular cells.[1,2] Also, IL-6 receptor polymorphisms have been identified in HIV-negative CD and are associated with increased soluble IL-6 receptor levels.[3]

DISCUSSION OF MANAGEMENT

Castleman Disease (CD) is a rare, benign disease of unknown aetiology, which is histologically characterised by angiofollicular lymph node hyperplasia most commonly in the mediastinum. CD can present as a localised mass first described by Castleman in 1954[4] or in a more aggressive multicentric form first reported by Gaba[5] also known as giant lymph node hyperplasia, angiomatous lymphoid hamartoma and follicular lymphoreticuloma. It has three histologic subtypes- hyaline vascular and plasma cellular and mixed. Most commonly occurs between 2nd and 4th decade. There is an equal incidence among males and females. The mediastinum is the most common site accounting for 60% of cases, the neck is involved in only 14% cases, more than 90% of neck lesions are unifocal. Usually, 5-10 cm in size presented as an ovoid neck mass. Clinically, Castleman disease is either unifocal or multifocal. Unifocal lesions have benign course cured by surgery and multicentric lesion are aggressive course, maybe fatal. Up to 30% lesions have malignant transformation, also transform into non-Hodgkin lymphoma or Kaposi sarcoma. Treatment for these lesion is chemotherapy or radiotherapy. It has been further subclassified into two types hyaline-vascular and plasma cell variants. Some may present with another mixed variant.[6] On ultrasonography, Castleman disease lesions appear as nonspecific, well-defined hypoechoic masses with intense peripheral vascular flow with small scattered foci centrally on colour Doppler. Konno et al have described a case of mesenteric Castleman disease characterised by a large feeding artery penetrating the nodal hilum and prominent arteries in the periphery identified on a Doppler sonogram. The evaluation of cervical and axillary Castleman diseases sonography remains useful in which the depiction of prominent peripheral vessels and penetrating feeding vessels on Doppler sonograms can suggest the diagnosis of this uncommon disease.[7] Castleman disease of the neck on CT scan has been described as well-circumscribed

homogeneous oval mass lesion, which is isodense to muscle with moderate to marked enhancement due to greater vascularity in the hyaline vascular type having a tendency to enhance more than the plasma cell type. On MRI of Castleman disease of the neck described as mass lesion, which is hypointense isointense to muscle on T1 weighted sequences, hyperintense oval mass on T2 weighted sequences.[8] Some authors have described the presence of linear hypointense signals on T2 weighted sequences in a stellate or arborising pattern, which is often not seen. They attributed these hypointense signals to perivascular lamellar fibrosis or sinus histiocytes and radial fibrosis and suggested that as an important diagnostic clue of Castleman disease. On PET findings are described as mild-to-moderate tracer uptake in unifocal, high tracer uptake in multicentric disease. Gallium-67 showing moderate tracer uptake. Best diagnostic clue on CT is solitary moderate to markedly enhancing mass considered as gold standard for diagnosis. However, biopsy requires for definitive diagnosis.

Summary

Although, most Castleman disease lesions typically appear as well-defined solitary hypoechoic neck masses on ultrasound, the findings are nonspecific. Further evaluation is to be done by other investing modalities like CT and MRI, which shows solitary homogenous well-circumscribed oval mass lesion with moderate to marked enhancement. Definitive diagnosis requires biopsy.

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