Indian Journal of Basic & Applied Medical Research; March 2013: Issue-6, Vol.-2, P. 582-584

"Solitary Castleman's Disease-At an unusual Site (Axilla) - A Case Report."

[1]Dr. Joshi Sneha R, [2]Dr. Tekwani Deepa T*

[1] Professor & HOD, Department Of Pathology,
MIMER Medical College , Talegaon Dabhade, Pune.
[2]Associate Professor, Department Of Pathology,
MIMER Medical College, Talegaon Dabhade, Pune.

ABSTRACT:

A 28 Years old female presented with a right axillary swelling in Talegaon General Hospital in April 2007. The patient was afebrile and on examination, had no organomegaly or any other lymphadenopathy. Her haematological , biochemical & radiological investigation did not reveal any abnormality. Clinically, the case was diagnosed as ?Lipoma/ ?Neurofibroma, which was followed by excision. A well-circumscribed specimen, measuring 8x8x3cm, soft to firm in consistency and grey in colour was received in the department of Pathology, MIMER Medical College, Talegaon Dabhade. The histopathological diagnosis of Castleman's Disease – Hyaline Vascular type (Solitary form) was offered. The solitary form of Castleman's Disease most commonly presents as a mass located in the mediastinum. This case is presented because of its occurrence in the axillary region – an unusual site.

Key words:- Axillary mass, Solitary form of Castleman's Disease, Hyaline-Vascular Type.

Background: Castleman's disease is a rare benign lymphoproliferative disorder of uncertain origin which most commonly involves the mediastinum but rarely affects the axilla[1]. It represents a morphologically distinct rare form of massive lymph node hyperplasia rather than a neoplasm or hamartoma[2]. Clinically, two forms of the disease exist: the localized or solitary form and the multicentric or systemic form[3]. Histologically, two distinct patterns have been described- the hyaline vascular type and the plasma cell type[2]. We report a case of Solitary Castleman's Disease –Hyaline Vascular type in a 28 year old female with an unusual presentation as an axillary mass.

CASE REPORT: A 28 year old female presented with a right axillary swelling. The swelling was non-tender, mobile and gradually increased in size to attain the present size of 9X8X4 cm over a period of 3 years. There were no complaints of fever and weight loss. The general examination of the patient did not reveal any any other specific positive finding except an axillary swelling. There was no hepatosplenomegaly.

Local examination revealed a firm swelling, measuring 9X8X4 cm in the right axillary region. The skin over the swelling appeared normal. Her haematological and biochemical investigations did not reveal any abnormality. Clinically, the case was diagnosed as a benign soft tissue tumor-? Lipoma/? Neurofibroma. Surgical excision of the mass was done and the specimen was sent to the Department of Pathology. Grossly, the mass was well circumscribed; encapsulated and measured 8X8X3cm.External surface was smooth and showed prominent blood vessels. [Fig.No.1]. The mass was firm in consistency and on cut section appeared solid gray with no areas of haemorrhage and necrosis. [Fig.No. 2].

Microscopic examination of the haematoxylin-eosin stained sections showed lymph node with marked follicular hyperplasia (Fig.No.3). The follicles were of varying size and showed prominent germinal centres.

At places the germinal centres showed vascular proliferation and hyalinization (regressive germinal centres). Moreover, there was a targetoid alignment of lymphocytes in the mantle zone of the hyalinized follicle with an occasional capillary penetrating radially into the germinal centres(Lollipop appearance) (Fig.No.4). Interfollicularstroma was prominent and showed lymphocytes, numerous hyperplastic vessels and hyaline change. The sinuses were absent. Considering the characteristic pathological features and the axillary location, the diagnosis of Solitary Castleman's Disease-Hyaline Vascular type (axilla) was given.

DISCUSSION: Synonyms include Angiofollicular lymph-node hyperplasia, Angiofollicular lymphoid hyperplasia, Giant lymph-node hyperplasia, Lymphoidhamartoma, Benign lymphoma or Follicular lymphoreticuloma.[3] Castleman's disease is a very rare pathology with unknown precise incidence. More than 400 cases of isolated case are reported in literature so far.[3] It occurs at any age, with a peak incidence in the third to fourth decade of life, but the multicentric form usually affects older individuals. Both types have equal male to female incidence.[5]The patient in the present case report was a 28 year old female. The disease is clinically classified into two major types: the localized or solitary form and the multicentric or systemic form.[3]

The most common site of the localized form is the mediastinum-70%, head and neck <10%. Additional sites of occurrence include the retroperitoneum, mesentery, vulva, pancreas and pelvis.[5]

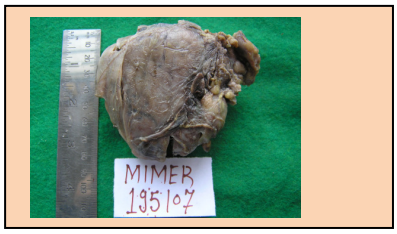
The incidence of solitary form of axillary Castleman's disease is 4%.[6] Grossly, it is known to be well-circumscribed and can measure even 15 cm or more in diameter. The cut surface does not show any necrosis.[2]In the present case report, the mass measured 8X8X3 cm, with no areas of necrosis on cut section.

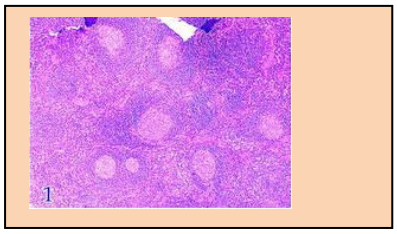
Histologically, two distinct patterns have been described-the hyaline vascular or angiofollicular type and plasma cell type. According to Keller et al., the hyaline vascular variant comprises 91% of Castleman's disease and mostly presents as a solitary lymphadenopathy[7]. This case also presented with a solitary axillary swelling.

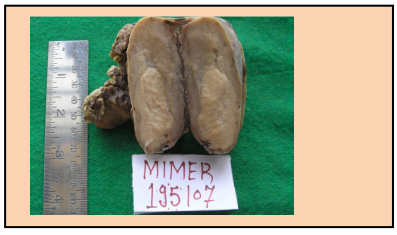
Microscopically, the hyaline vascular type shows increased number of scattered large follicles, many of which with hyalinization and marked vascular proliferation of the germinal centres.[2] Often capillaries penetrating follicles give a "lollipop" appearance.There is a tight concentric layering of lymphocytes at the periphery of the follicle, resulting in an "onion-skin", "targetoid" or "stadium seating" appearance.[8] The interfollicular area shows marked proliferation of post capillary venules and presence of plasma cells, eosinophils, immunoblast and monocytes.Sinuses are characteristically absent.[2]All these characteristic histological features of Castleman's disease were noted in the present case. The etiology is unknown, the two main hypothesis being abnormal immune response and viral infection.[2] Human herpes virus 8 and interleukin 6 are regarded to be linked to the pathogenesis.[9] Dysplastic or atypical follicular dendritic cells positive for CD21 and CD 35 have frequently been described in the hyalinizedcentre and can even show monoclonality, yet their role in the pathogenesis is unclear.[2,8,] Solitary Castleman's disease follows a benign course, and in most cases, surgical removal of the affected lymph node is curative as recurrence is not expected.[10] The patient of our case report did not reveal any recurrence.

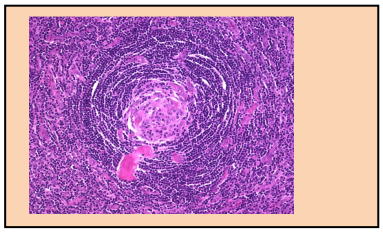
References:

1) Seo BK, Oh YW, Cho KR, Lee NJ, Kim JH, Kim IS, Cho SJ, Bae JW .Imaging findings of Castleman's Disease Localized in the Axilla: A Case Report. Korean J Radiol. 2002 Apr-Jun; 3(2):136- 139.
2) Rosai J. Lymph nodes. Chapter 21. In: Rosai J, editor. Ackerman′s Surgical Pathology. 9th ed. St Louis: Mosby; 2003. p. 1877-2017.
3) Sarrot-Reynauld F. Castleman′s disease. In: Orphanet encyclopedia [Online]. [1999] Available from: http://www.orpha.net/data/patho/GB/uk-castleman.pdf .[last cited on 2001 Aug].
4) Castleman B, Iverson L, Mendez VP. Localized mediastinal lymph node hyperplasia resembling thymoma. Cancer 1956; 9:822-830.
5) S.Raniga, C.Shah, A. Shrivastava, P. Amin, P. Patel. Doppler Findings In Castleman Disease-A Rare Case. Ind J Radiol Imag 2006, 16(1): 127-130.
6) Arnab Ghosh, Seema V. Pradhan, O.P.Talwar. Castleman′s disease-hyaline vascular type-clinical,cytological and histological features with review of literature. IJPM 2010;53(2):244-247.
7) Keller AR, Hochholzer L, Castleman B. Hyaline-vascular and plasma-cell types of giant lymph node hyperplasia of the mediastinum and other locations. Cancer. 1972;29:670-681.
8) Roshong-Denk SL, Bohman SL, Booth RL. A 53-year-old white man with right- sided supraclavicular lymphadenopathy. Arch Pathol Lab Med 2005;129:945-6.
9) Mallik MK, Kapila K, Das DK, Haji BE, Anim JT. Cytomorphology of hyaline- vascular Castleman′s disease: a diagnostic challenge. Cytopathology 2007; 18:168-74.
10) Biswas B. Angiofollicular lymph node hyperplasia (Castleman′s disease) - A case report. IJTCVS 2004;20:201-3.









Date of Submission: 22 December 2012
Date of Provisional acceptance: 08 January 2013
Date of Final acceptance: 16 February 2013
Date of Publication: 05 March 2013
Source of Support: Nil Conflict of Interest: Nil