Case Report

A rare case report of Castleman's disease and review of literature

Atul Kothari[1*], Smita Patel[2], Jigar Shah[3], S S Goswami[4]

[1]Resident Doctor, [2]Professor, [3]Assistant Professor, [4]Professor
Department of Pathology, SBKS MI and RC, Sumandeep Vidyapeeth, Vadodara, Gujarat, India
[*]Corresponding author email: **ak32atul@gmail.com**

International Archives of Integrated Medicine, Vol. 9, Issue 10, October, 2022.
Available online at http://iaimjournal.com/
ISSN: 2394-0026 (P) **ISSN: 2394-0034 (O)**
Received on: 24-9-2022 **Accepted on:** 1-10-2022
Source of support: Nil **Conflict of interest:** None declared.

How to cite this article: Atul Kothari, Smita Patel, Jigar Shah, S S Goswami. A rare case report of Castleman's disease and review of literature. IAIM, 2022; 9(10): 39-42.

Abstract

Castleman's disease (CD) is a rare lymphoproliferative disorder, described for the first time by Castleman, et al. in 1954 in a single case then in 1956 in a group of 13 patients with localized benign lymphadenopathy. The incidence of CD is unknown and can occur at any age, however it is mainly reported in adults in the literature with a slight feminine predominance (60%). The majority of the previously reported cases of CD in the neck were of the hyaline vascular type and the most common sign was an asymptomatic neck mass. In the present case the patient is having unicentric CD and histologically it is Mixed Type. We are reporting this case because of its rarity.

Key words

Castleman's disease, Cervical, Lymphadenopathy.

Introduction

Castleman's disease (CD) is an uncommon clinicopathological entity characterized by non-neoplastic lymph node hypertrophy and histologically characterized by angiofollicular lymph node hyperplasia. The disease was first described by Dr. Benjamin Castleman in a single case in 1954 [1], followed by a small cases series in 1956 [2]. Clinically the disease has 2 forms. One is localized as first described by Castleman, which is more common and other one is multicentric disease (MCD) with involvement of several sites, which was first described by Gaba, et al. in 1972 [3]. The majority of the previously reported cases of CD in the neck were of the hyaline vascular type and the most common sign was an asymptomatic neck mass [4]. In the present case the patient is having unicentric CD and histologically it is Mixed Type. We are reporting this case because of its rarity.

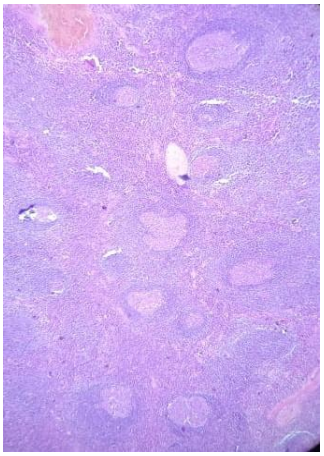
Case report

A 36 year old female patient presented with history of swelling on left sided submandibular region of the neck for two years in the OPD of Department of General Surgery at Dhiraj General Hospital, Waghodia. The painless swelling was gradually increasing in size. There was no history of constitutional symptoms like fever, weight loss and night sweats. There was no history of pain in the throat, dysphagia, dyspnea change of voice and exposure to tuberculosis either. General examination of the patient was normal. Neck examination showed a firm, non-warm, non-tender, non-reducible swelling, in the submandibular region of neck on the left side measuring 8×5cms in its greatest dimension which was freely mobile in all directions. There was no movement on deglutition or protrusion of tongue. There were no generalized lymphadenopathies, hepatomegaly or splenomegaly. All Hematological, Serological and Biochemical investigations were within normal limit.ESR were raised at 43mm after 1 hour. Fine needle aspiration cytology (FNAC) was advised by the clinicians and patient referred to cytopathology laboratory. FNAC was performed with all precautions after taking consent. [5-11].

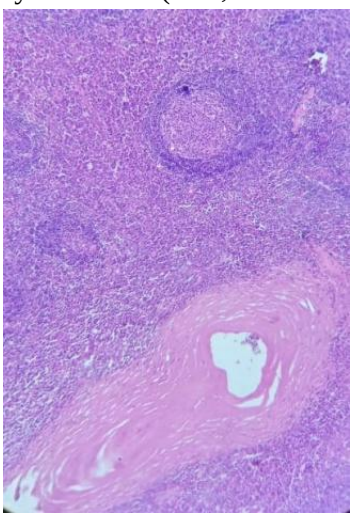
The FNAC smears showed richly cellular smears with medium sized lymphoid cells, groups of histiocytic cells and immunoblasts. Cytopathologist advised Excision biopsy and detailed histopathological examination. A surgical excision of the mass was carried out under general anesthesia and specimen sent to Histopathology section of the department of Pathology. On gross examination 6×4×2 cm soft tissue mass was received with grayish white cut surface (**Photograph – 1**). On microscopic examination, the sections showed large follicles scattered in a mass of lymphoid tissue. The follicles showed vascular proliferation and hyalinization. There is a tight concentric layering of lymphocytes at the periphery of the follicles, resulting in onion-skin appearance. The inter follicular stroma is also prominent and infiltrated with predominantly plasma cells, lymphocytes and eosinophils accompanied by numerous russel bodies (**Photograph - 2, 3, 4**). From over all histopathological examination findings final diagnosis was made as Castle- man disease-Mixed Type (Unicentric).

Photograph – 1: Greyish white soft tissue.

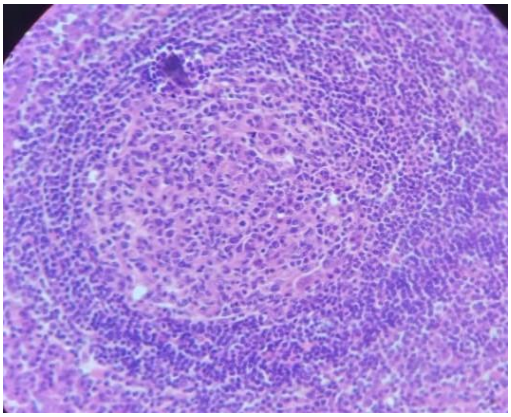
Photograph – 2: Large follicles scattered in a mass of lymphoid tissue (4X, H&E stain).



Photograph – 3: Vascular proliferation and hyalinization (10X, H&E stain).



Photograph – 4: Tight concentric layering of lymphocytes at the periphery of the follicles, resulting in onion-skin appearance (20X, H&E stain).



Discussion

CD is a rare lymphoproliferative disorder, described for the first time by Castleman, et al. in 1954 in a single case then in 1956 in a group of 13 patients with localized benign lymphadenopathy. The Synonyms of CD are Angiofollicular lymph node hyperplasia, Angiofollicular lymphoid hyperplasia, Giant lymph node hyperplasia, lymphoid hamartoma and Benign lymphoma. The incidence of CD is unknown and can occur at any age, however it is mainly reported in adults in the literature with a slight feminine predominance (60 %). Even though the exact pathogenesis of CD is vague, studies have indicated that the occurrence of CD may be related to the infection of human herpes virus-8 (HHV-8) or human immunodeficiency virus (HIV), immune dysfunction, and excessive production of interleukin-6 (IL-6). Histologically there are three different types of CD: hyaline-vascular type, plasma cell type and mixed variant type [4, 12]. Hyaline-vascular type is the most common clinical finding (90 %) characterized by follicular hyperplasia with regressed germinal centers and important vascular proliferation. Plasma cell type is characterized by Russell bodies and exhibits larger lymphoreticular nodules and fewer hyalinized blood vessels compared with the hyaline-vascular type. Mixed variant type is a rare variant and exhibits features of hyaline-vascular and plasma cell type [4]. In our case we had found all features of mixed type of CD.

CD often shows well-defined, mildly hypodense or isodense, homogeneous nodules or masses on different imaging modalities (CT/MR), and intermediate and marked enhancement on contrast-enhanced CT/MR images. The hypertrophy of blood vessels is also considered as valuable features [13]. The calcification in affected lesions is common and is more commonly observed in hyaline vascular variant unicentric CD [14]. Fine needle aspiration cytology (FNAC) findings may not always be conclusive as in our case. CT and MRI are usually of great value in the case prognosis. Surgical resection is considered the cornerstone of radical treatment for unicentric CD and is the most widely accepted therapy in the literature therefore the upfront excision decision was taken in our case for diagnostic and radical concerns. Diagnosis is confirmed by histopathological assessment of the lymph node biopsy sent postoperative [12, 15].

If surgical removal isn't possible, in cases that the lymph node is difficult to get to, radiation therapy [16] may be an effective way to destroy the affected tissue. In the plasma cell and multicentric types, corticosteroids [17], chemotherapeutic drugs, monoclonal antibodies and immunomodulators may be used [18]. This approach is usually reserved for patients refusing surgery or having multicentric disease. Long term follow up of patients of Castleman's disease shows very low or nil recurrence in cases which have been successfully treated. However there should be a constant look out for malignant sequel [19].

Conclusion

Cervical lymph nodes are involved by Castleman's disease and may be confused with other common causes of cervical lymphadenopathy like tuberculosis and nodal secondaries. This entity is alarming for all pathologists because of its rarity. One should

keep in mind CD while diagnosing any swelling in neck. Histopathology is the gold standard for the final diagnosis.

References

1. B. Castleman, V.W. Towne. Case records of the Massachusetts General Hospital; weekly clinicopathological exercises; founded by Richard C. Cabot. N. Engl. J. Med., 1954; 251(10): 396–400.
2. B. Castleman, L. Iverson, V.P. Menendez. Localized mediastinal lymph node hyperplasia resembling thymoma. Cancer, 1956; 9(4): 822–830.
3. Gaba AR, Stein RS, Sweet DL, Variakojis D. Multicentric giant lymph node hyperplasia. Am J Clin Pathol., 1978; 69: 86–90.
4. L. Jiang, L.Y. Zhao, Y. Liu, Y.F. Zhao. Castleman´s disease of the neck: report of four cases with unusual presentations, J. Oral Maxillofac. Surg., 2011; 69(4): 1094–1099.
5. Gunvanti Rathod, Pragnesh Parmar. Fine needle aspiration cytology of swellings of head and neck region. Indian Journal of Medical Sciences, 2012; 66: 49-54.
6. needleaspiration cytology in cervical lymphadenopathy – A one year study. International Journal of Medical and Pharmaceutical Sciences, 2014; 4(5): 1-8.
7. Rathod GB, Ghadiya V, Shinde P, Tandan RK. Pleomorphic sarcoma in 60 years old male – A case report. International Journal of Current Microbiology and Applied Sciences, 2014; 3(8): 510-517.
8. Gunvanti Rathod, Pragnesh Parmar, Sangita Rathod, Ashish Parikh. Suprascapular malignant fibrous histiocytoma – A case report. Discovery, 2014; 12(31): 50-53.
9. Rathod GB, Goyal R, Bhimani RK, Goswami SS. Metaplastic carcinoma of breast in 65 years old female - A case report. Medical Science, 2014; 10(39): 77-81.
10. Disha Singla, Gunvanti Rathod. Cytodiagnosis of renal cell carcinoma – A case report. IAIM, 2015; 2(2): 133-137.
11. Mobeen Alwani, Gunvanti B. Rathod. Diagnosis of anaplastic thyroid carcinoma on fine needle aspiration cytology - A rare case report. IAIM, 2015; 2(3): 183-187.
12. G.M. Chronowski, C.S. Ha, R.B. Wilder, F. Cabanillas, J. Manning, J.D. Cox, Treatment of unicentric and multicentric Castleman disease and the role of radiotherapy, Cancer, 2001; 92(3): 670–676.
13. N. Talat, A.P. Belgaumkar, K.M. Schulte. Surgery in Castleman´s disease: a systematic review of 404 published cases, Ann. Surg., 2012; 255(4): 677–684.
14. A. Sudha, N. Vivekanand. Cytologic picture of Castleman´s disease: a report of two cases, J. Cytol. Indian Acad. Cytol., 2010; 27(4): 152–154.
15. S. Iyer, M.I. Bhatti, M. Halliday. Castleman´s disease - A case report. Int. J. Surg. Case Rep., 2010; 1(3): 25–26.
16. Bowne WB, Lewis JJ, Filippa DA, Niesvizky R, Brooks AD, Burt ME, et al. The management of unicentric and multicentric Castleman's disease. Cancer, 1999; 85: 706-17.
17. Summerfield GP, Taylor W, Bellingham AJ, Goldsmith HJ. Hyaline-vascular variant of angiofollicular lymph node hyperplasia with systemic manifestations and response to corticosteroids. J Clin Pathol., 1997; 36: 1005-11.
18. Tamayo M, Gonzalez C, Majado MJ, Candel R, Ramos J. Long-term complete remission after interferon treatment in a case of multicentric Castleman's disease. Am J Hematol., 1995; 49: 359-60.