JIDAM
eISSN 2582 - 0559
"An Official Journal of IDA - Madras Branch"©2019.
Available online

CASE REPORT

CASTLEMAN'S DISEASE OF THE MASSETER & SUBMENTAL REGION – A RARE CASE REPORT

Madhumitha.M, Rekha.M*, Dineshkumar.T**.

Department of Pediatric and Preventive Dentistry
*Department of Oral and Maxillofacial Surgery
**Department of Oral Anatomy and Oral Pathology

To access & Cite this article
Website: www. jidam.idamadras.com

Address for Correspondence:

Dr. M. Madhumitha, MDS,
D225, Manchester Grand Apartments,
M.G Road, Avarampalayam,
Coimbatore, Tamilnadu, India.
E-mail : madhumithakarthik@gmail.com

Received : 28.12.2018
Accepted : 26.02.2019
Published : 26.03.2019

ABSTRACT

Castleman's disease is a rare clinical entity presenting either as localised benign tumours affecting chiefly the lymph nodes or as widespread systemic disease. The incidence of castle man's disease occurring in the head and neck region is very rare and hence it is not much known. The disease is thought to be due to over production of Interleukin 6 or due to hyper responsiveness to Interleukin 6. Human herpes virus type 8 (HHV -8) is also proposed to be associated with castle man's disease. The localized type is treated by simple surgical excision. Though recurrence is rare, the plasma cell type requires regular follow up. The multicentric type often requires aggressive systemic therapy with corticosteroids and has poor prognosis. In some patients, the multicentric disease has been associated with HIV infection. Here, we report a case of castle man's disease in a 32 year old male patient involving the masseter and sub mental region to emphasis the importance of including this disease in the differential diagnosis of swellings in the head and neck region.

KEY WORDS: Castleman's disease, masseter, sub mental region

INTRODUCTION:

Castleman's disease is a rare disorder, presenting clinically as benign tumors. It either presents as a localized disease like a mediastinal mass[1], or like giant lymph node enlargements[2] or as a systemic disease. The localized type / solitary type is usually asymptomatic[3], self-limiting and present as a mass or swelling in the chest, stomach and rarely in the head & neck region. The systemic / multicentric type is characterized by fever with chills, generalized lymphadenpathy, anemia and hepatosplenomegaly and is highly aggressive frequently resulting in death despite aggressive management. Usually the growths are nothing but lymphoid hamartomas [2].

CASE REPORT:

A 32 year old male presented to the department of OMFS with a history of progressive painless swelling in the left masseteric region for the past one month. He gave a history of similar swelling in the right and left lateral aspect of the neck and was treated by a General medical practitioner with medications. He had other constitutional symptoms such as weight loss and weakness including loss of appetite for the last 2 months. On general medical examination, there was no associated generalized lymphadenopathy. On examination, the swelling in the left sub-masseteric region (Fig 1) was non-tender, firm in consistency with ill-defined margins. The overlying skin was normal. Examination of oral cavity, pharynx, and Larynx did not reveal any possible focus of infection or pathology. As the lesion became firm on clenching

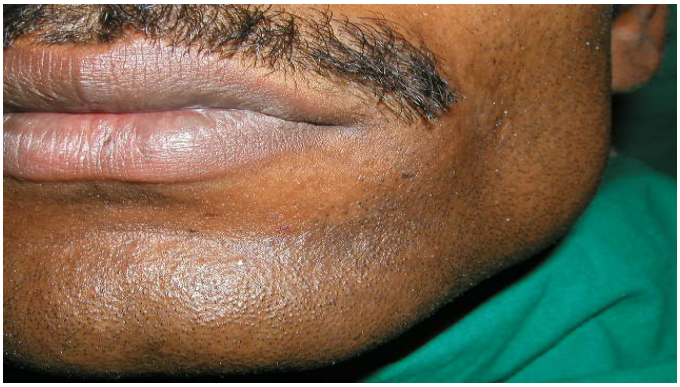

Fig 1: Swelling in the left sub-masseteric region which was non-tender, firm in consistency with ill-defined margins.

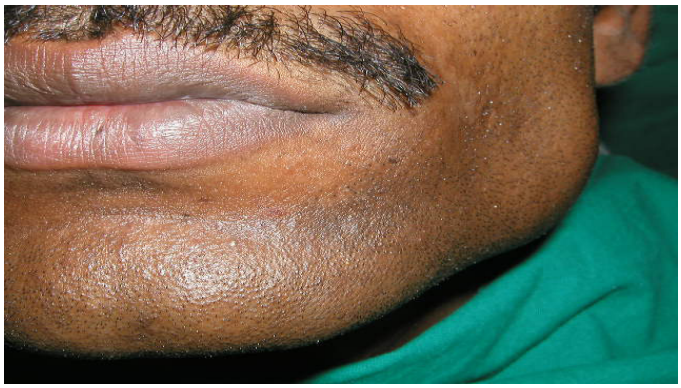

Fig 2: The swelling became firm on clenching

(Fig 2), a suspected clinical diagnosis of massetric hypertrophy was made. OPG and PA Cephalogram were normal. All routine blood investigations were done & there was increase in counts of lymphocytes & eosinophils. Chest x- ray and ESR were normal. A USG study of the masseter region, showed increased contractions of the masseter

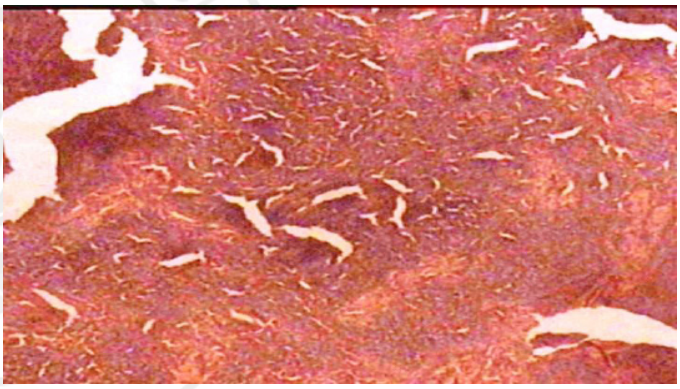

Fig 3: The histopathology of the given specimen shows hyperplastic vessels, a mixture of plasma cells, eosinophils and proliferation of immunoblasts.

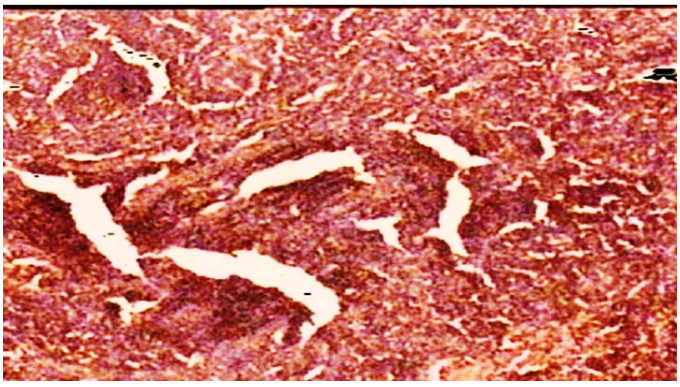

Fig 4: The histopathology shows proliferation of lymphoid follicles with hyalinized vessels and coaxial layers of lymphocytes in the periphery.

muscle, but failed to show any specific mass. Both the parotid glands appeared normal. After three days without any treatment the swelling disappeared and the masseteric area appeared normal to our surprise. The patient was kept under observation. A month later patient reported back with a similar swelling in the submental region. On clinical examination, the swelling was warm & tender. Bilateral submandibular lymph nodes were also palpable. Mantoux test was positive (11mm). HIV & HBsAg screening tests were negative. The USG abdomen showed no significant findings. Lymph node biopsy was done and was sent for histopathological examination. The histopathological examination of the excised lymph node revealed hyperplastic vessels & an admixture of plasma cells, eosinophils & immunoblasts (Fig 3,4) in the lymphoid tissue showing a picture of Castleman's disease of Hyaline vascular type. Even though treatment was suggested, the patient failed to attend further appointments. However one year after, the patient reported back to the department and was found to be asymptomatic without any problems.

DISCUSSION:

Castleman et al described this disease as localized lymph node masses occurring in the mediastinal region[1]. The disease usually occurs more often in young individuals with males and females being equally affected. The commonest site is mediastinum followed by other nodal sites such as retroperitoneal, paraaortic, auxiliary, and cervical. Cervical node [3, 4] is the least commonly involved site. It was also reported in muscle, lung, larynx, parotid[5] and pancreas. In the maxillofacial region, it was reported in orbits[6] and buccal mucosa[7] also.

The overproduction of interleukin-6 (IL-6)[8] or hyper-responsiveness to IL-6 is suggested to be responsible for the pathogenesis of castle man disease. The genome of HHV-8 harbors a viral analogue of the IL-6 gene and hence Human herpes virus type 8 (HHV-8) is associated with Castleman disease[9]. Increased levels of tumor necrosis factor-β (TNF-β), interferon-γ (IF-γ), vascular endothelial growth factor (VEGF) and macrophage colony-stimulating factor are also proposed to be responsible for the pathogenesis of the disease process. Profound immunodeficiency in multicentric Castleman Disease might be explained by apoptosis of T cells[10].

There are four histopathological variants of this disease

(1) Hyaline vascular type: This is the most common histopathological pattern of the disease accounting for 80-90% of the cases. This type usually presents as a single, enlarged lymph node. The excised lymph node shows abnormal lymphoid follicles with atropic, hyalinized follicular centers and small lymphocytes in a concentric arrangement and penetrating blood vessels.

(2) Plasma cell type: less common than hyaline vascular type. The histopathology shows presence of plasma cells in the interfollicular areas. In contrast to the atrophic follicular centers seen in hyaline-vascular type, the follicular centers are usually enlarged and hyperplastic with absence of paracortical hypervascularity and onionskin mantle zones.

(3) Transitional type: occupies intermediate zone between hyaline vascular type and plasma cell type.

(4) Stromal rich type: contains enlarged interfollicular zone. The localized type can be cured by simple surgical excision regardless of histological type. The excision is both diagnostic and therapeutic. The plasma cell type lesion is reported to have more recurrences than the hyaline vascular type; hence, follow-up is required in plasma cell type. The multicentric disease10 has poor prognosis and requires aggressive systemic therapy. The multicentric form is treated with corticosteroids that inhibit IL-6 production and combination chemotherapy aimed at providing complete remission. But the optimal regimen still needs to be standardized. CHOP (Cyclophosphamide, Doxorubicin, Vincristine and Prednisone) regimen is indicated for successful treatment of refractory cases.

In few cases, the multicentric disease is associated with HIV infection, linked to human herpes virus type 8 (HHV-8), also known as Kaposi sarcoma-associated virus. There is a high incidence of malignancies, (lymphoma and Kaposi's sarcoma) in about 37% of patients with multicentric disease.

Furthermore, Hodgkin's lymphoma can occur in cervical Castleman's disease. Although complete surgical excision of localized castleman's disease is the treatment of choice, a standard lymphoma workup and management strategy is needed to prevent management of lymphomas developing in these lesions[4].

CONCLUSION:

It is important to know this poorly understood disease, which can occur at any age and resembles other common lesions such as tuberculosis and sarcoidosis. Any suspected lymphadenopathy should be investigated properly and possible biopsy of the lymph node should be done to rule out these types of rare lesions.

FINANCIAL SUPPORT AND SPONSORSHIP:

Nil

CONFLICT OF INTEREST :

There is no conflict of interest

REFERENCES:

1. Castleman B, Iverson L, Menendez PV. Localised mediastinal lymphnode hyperplasia resembling thymoma. Cancer 1954;9: 822-30.
2. Penfold CN, Cottrell BJ, Neonatal giant lymph node hyperplasia (Castleman's disease) presenting in the head and neck.Br J Oral Maxillofac Surg. 1991 Apr; 29(2): 110-1
3. Calli C, Oncel S, Pinar E, Guclu E, Calli A, Apaydin M. Castleman's disease of the cervical region: a case report Kulak Burun Bogaz Ihtis Derg. 2005; 14(1-2):35-8.
4. Frank DK, Charney D, Kashani A. Plasma cell variant of Castleman's disease occurring concurrently with Hodgkin's disease in the neck .John Wiley & Sons, Inc. Head Neck 23(2): 166-169, 2001.
5. Leocata P, Corbacelli A, Chiominto A, Cutilli T, Belli L, Ventura L. Unicentric angiofollicular hyperplasia (Castleman's disease) of the parotid-a case report. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1996; 81: 328-32.
6. Keun-Sung Park1, You-Jin Choi1 and Kyu-Sang Hyaline-vascular type Castleman's disease involving both orbits Acta Ophthalmologica Scandinavia, 2002 October Volume 80 Page 537
7. Satoshi Maruyama Natsuko Hao Jun Cheng , Kazuhito Horino , Makoto Ohnishi , Masaya Fukushi , Masahiro Fujii and Takashi Saku Castleman's Disease of the Buccal mucosa – Report of a case and review of the Literature of Head and Neck cases. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2002; 93:305-10
8. Beck JT, Hsu S, Wijdenes J et al. (1994): Alleviation of systemic manifestations of Castleman's disease by monoclonal anti-interleukin-6 antibody. N Engl J Med 330: 602–605.
9. Parravinci C, Corbellino M, Paulli M et al. (1997): Expression of a virus derived cytokine, KSHV vIL-6, in HIV-seronegative Castleman's disease. Am J Pathol 151: 1517–1522.
10. Ishiyama T, Koike M, Fukuchi K et al. (1996): Apoptosis of T cells in multicentric Castleman's disease. Clin Immunol Immunopathol 79: 3.