

Castleman's Disease of Submandibular Region: A Rare Case Report

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Abstract: Castleman's disease is a non neoplastic condition of the lymphnode seen most commonly in mediastenal region. It can present as either unicentric or multicentric disease. Here we report one interesting case of Castleman's disease in a male patient occurring in submandibular region, which is very rare location of presentation. The case is further more interesting as it emphasises the importance of histopathology which is the key to the diagnosis.

Keywords: Castleman's Disease, submandibular region, Hyaline vascular type.

INTRODUCTION

Castleman's disease was first described by Benjamin Castleman in a series of 14 cases in 1956 [1]. The disease has a rare occurrence and the etiology is unknown. The most common location is mediastinum and neck is a very rare site of presentation. Patients present with asymptomatic lymphadenopathy, but systemic symptoms are known to occur in cases with multicentric disease and plasma cell rich variant.

We here present a case of submandibular, unicentric Castleman's disease in a male patient.

CASE REPORT

A 47 year old male patient presented to the surgical outpatient department with an asymptomatic swelling in the submandibular region. The swelling was slowly increasing in size over a period of one and a half years. There was no other associated disease and the past medical history was unremarkable. On examination the mass was firm well circumscribed located in the submandibular region and not mobile. There were no other swellings noted. General examination was otherwise insignificant. The basic laboratory investigations like complete blood picture (CBP) and serum biochemistry were within normal limits. HIV test was non reactive. Chest X ray didn't reveal any mediastinal lesion. Ultrasound examination of the neck region revealed a well circumscribed hypo-echoic area in the submandibular region. Fine Needle Aspiration Cytology (FNAC) showed smears composed of polymorphous population of lymphoid cells and occasional histiocytes (Fig 1). A diagnosis of non specific reactive lymphadenitis was offered on cytology. But as the swelling did not subside with a course of

antibiotics, the mass was excised and sent for histopathological examination (HPE). At the department of pathology, we received a nodular soft tissue mass measuring 3X 2 X 1 cm. Cut surface was solid grey white and homogenous. The tissue was routinely fixed and processed and stained with haematoxylin & eosin. Microscopy revealed complete effacement of lymph node structure. The capsule was thickened at few places and there was obliteration of the sub-capsular sinus. Under the capsule, there was proliferation of lymphoid follicles with extensive areas of hyalinisation giving a characteristic onion peel appearance (Fig. 2, 3). There were also numerous interfollicular proliferating vessels along with scattered Eosinophils seen (Fig. 4). A diagnosis of Hyaline vascular type of Castleman's disease was made based on histology.

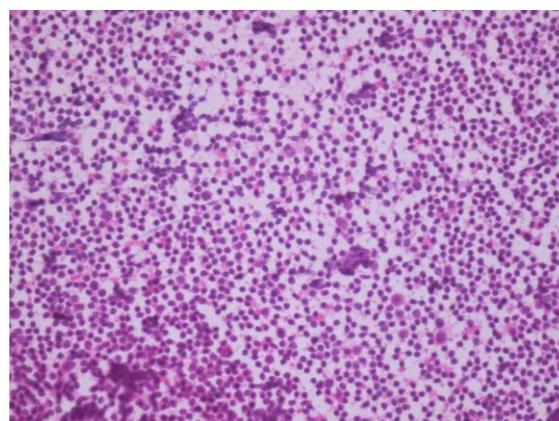


Fig. 1: Cytosmears showing polymorphous population of Lymphoid cells

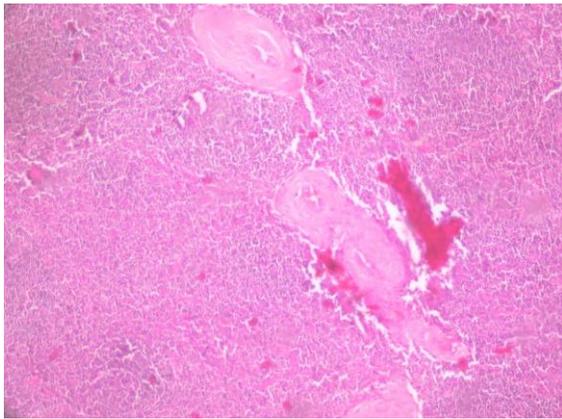


Fig. 2: Proliferating lymphoid Follicles and blood vessels

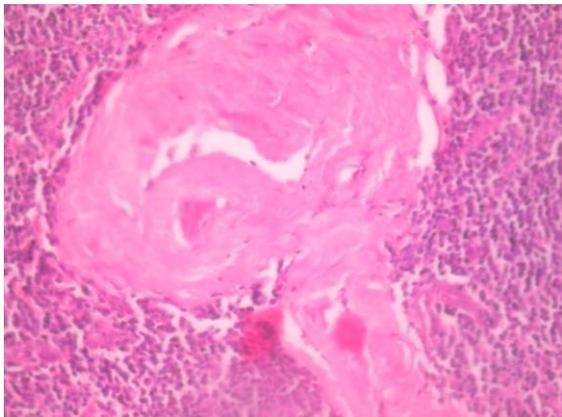


Fig. 3: Hyalinised stroma (onion peel appearance)

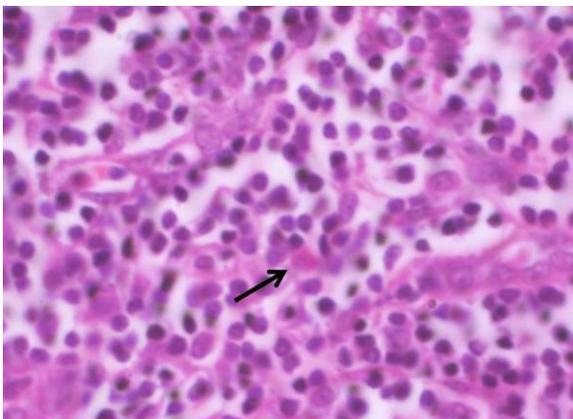


Fig. 4: Scattered Eosinophils (arrow)

DISCUSSION

Castleman disease is a non neoplastic enlargement of lymphoid tissue. It was first described by Dr. Benjamin Castleman in 1956. Synonyms include Angiolymphoid hyperplasia, Giant lymphnode hyperplasia, lymphnode hamartoma and benign lymphoma [2]. The etiology of the disease is unknown and any age group can be affected. Histologically, it can be Hyaline vascular type (80-90%), Plasmacytic type (10-20%) or rarely Mixed type [3]. Hyaline vascular type is characterised by proliferation of hyalinised follicles with proliferating vessels which gives a classic

onion peel appearance on microscopy. There is also infiltration with numerous Eosinophils in most of the cases. The Plasmacytic variant is characterised by hyperplastic germinal centres with extensive infiltration of plasma cells along with vascular proliferation. The proliferation of plasma cells is thought to be responsible for the systemic symptoms seen in these cases. These plasma cells are known to produce Interleukins which result in systemic manifestations like fever, raised ESR, weight loss etc., More over this variant is also seen to be associated with multicentric form of disease and diffuse organ involvement [3]. Mixed type is characterised by features of both the types of disease. Based on the nature of presentation it can be either unicentric or multicentric type. Most commonly multicentric disease is associated with HIV and Kaposi sarcoma (KS-HHV 8) [4]. Most common location is mediastinum(60%), other places with lymph-nodes can also be effected like retroperitoneum (11%), axilla (4%) etc. Head and neck region is effected in 14% of cases [5]. Unicentric form of disease has better prognosis and patients remain disease free for longer duration of intervals, the treatment of choice being simple resection. Multicentric form can also be associated with other systemic manifestations like anemia, nephrotic syndrome, vascular neoplasms, increased ESR, hypergammaglobulinemia etc. These patients need chemotherapy along with symptomatic treatment. Most patients with multicentric form of disease tend to develop complications like lymphoma during later stage of disease and thus require regular follow up [6, 7].

The present case is a 47 year male patient who presented with asymptomatic swelling of submandibular region. There were no other associated lesions and serology was non reactive for HIV. Cytology was performed initially and in view of the presence of mature lymphocytes alongwith a few centrocytes and centroblasts with histiocytes , a diagnosis of non specific reactive lymphadenitis was offered. With the lesion not responding to the routine treatment, the mass was excised. The histomorphology revealed proliferating follicles, hyalinised stroma along with interfollicular proliferation of blood vessels, features consistent with with Hyaline Vascular type of Castleman disease.

Following resection the patient is symptom free without any recurrence till date (more than 6 months). Thus, non invasive or minimally invasive techniques like radiology or aspiration cytology are rarely helpful in diagnosis marking histopathology as the key to diagnosis.

CONCLUSION

So we conclude that given the rarity of occurrence especially in head and neck regions and also association with HIV and the differential treatment modalities of the Hyaline vascular and Plasmacytic

type, accurate diagnosis by histopathology of Castleman's disease and the type is very important.

REFERENCES

1. Castleman B, Iverson I, Menendez VP; Localized mediastinal lymph-node hyperplasia resembling thymoma. *Cancer*, 1956; 9(4): 822-830.
2. Zhong LP, Wang LZ, Ji T, Hu YH, Hu YJ, Ye WM, Zhang CP; Clinical analysis of Castleman disease (hyaline vascular type) in parotid and neck region. *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology*, 2010; 109(3): 432-440.
3. Keller AR, Hochholzer L; Hyaline-vascular and plasma-cell types of giant lymph node hyperplasia of the mediastinum and other locations. *Cancer*, 1972; 29(3): 670-683.
4. Dupin N, Diss TL, Kellam P, Tulliez M, Du MQ, Sicard D, Weiss RA, Isaacson PG, Boshoff C; HHV-8 is associated with a plasmablastic variant of Castleman disease that is linked to HHV-8-positive plasmablastic lymphoma. *Blood*, 2000; 95(4): 1406-1412.
5. Gaba AR, Stein RS, Sweet DL, Variakojis D; Multicentric giant lymph node hyperplasia. *Am J Clin Pathol*, 1978; 69: 86-90.
6. Park JH, Lee SW, Koh YW; Castleman disease of the parotid gland in childhood: an unusual entity. *Auris Nasus Larynx*, 2008; 35(3): 451-454.
7. Somdas MA, Ketenci I, Bicer S, Senturk M, Guney E; Castleman's disease as an unusual neck mass: case report. *Ann Otol Rhinol Laryngol*, 2004; 113: 459-61.