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Two Rare Cases of Castleman Disease with Review of Literature

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Abstract: *Castleman disease is a very rare entity and Castleman disease involving the cervical region is even more rarer [1]. In this case series we have reported two cases of atypical neck mass of uncertain cytological diagnosis and not responding to conservative management. These two patients underwent excisional biopsy and the histopathological result came out to be Castleman Disease. Both these patients are symptom free till resent follow up of 5 and 9 months respectively with no radiological and clinical recurrence and are still under regular follow-up.*

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I. Introduction

Castleman disease is a rare clinicopathological disease which is included in the differential diagnosis of any lymphoproliferative disease [2]. Although not considered a malignant disease, one variety (Multicentric type) can later lead to the development of Non-Hodgkin's lymphoma [3]. Here we have document two cases of atypical neck swellingwhich after excisional biopsy were confirmed to be Castleman Disease.

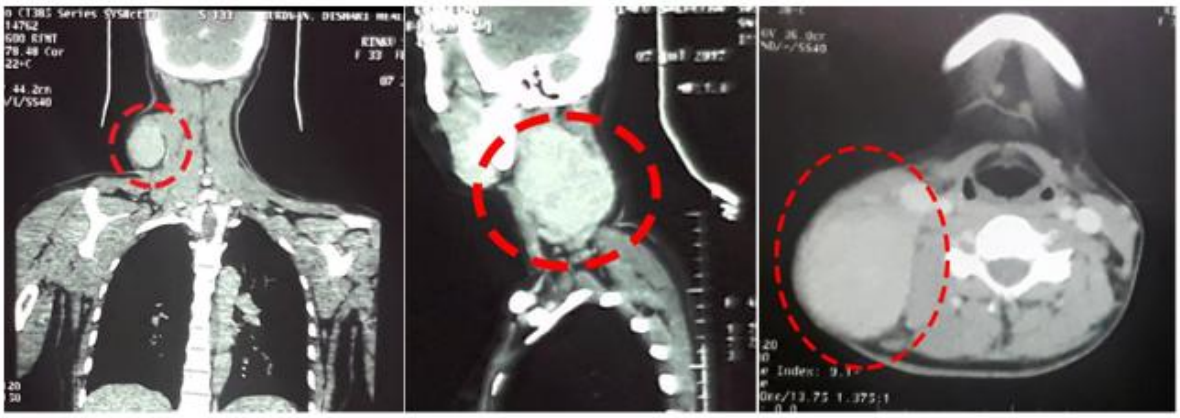
II. Case reports

CASE 1

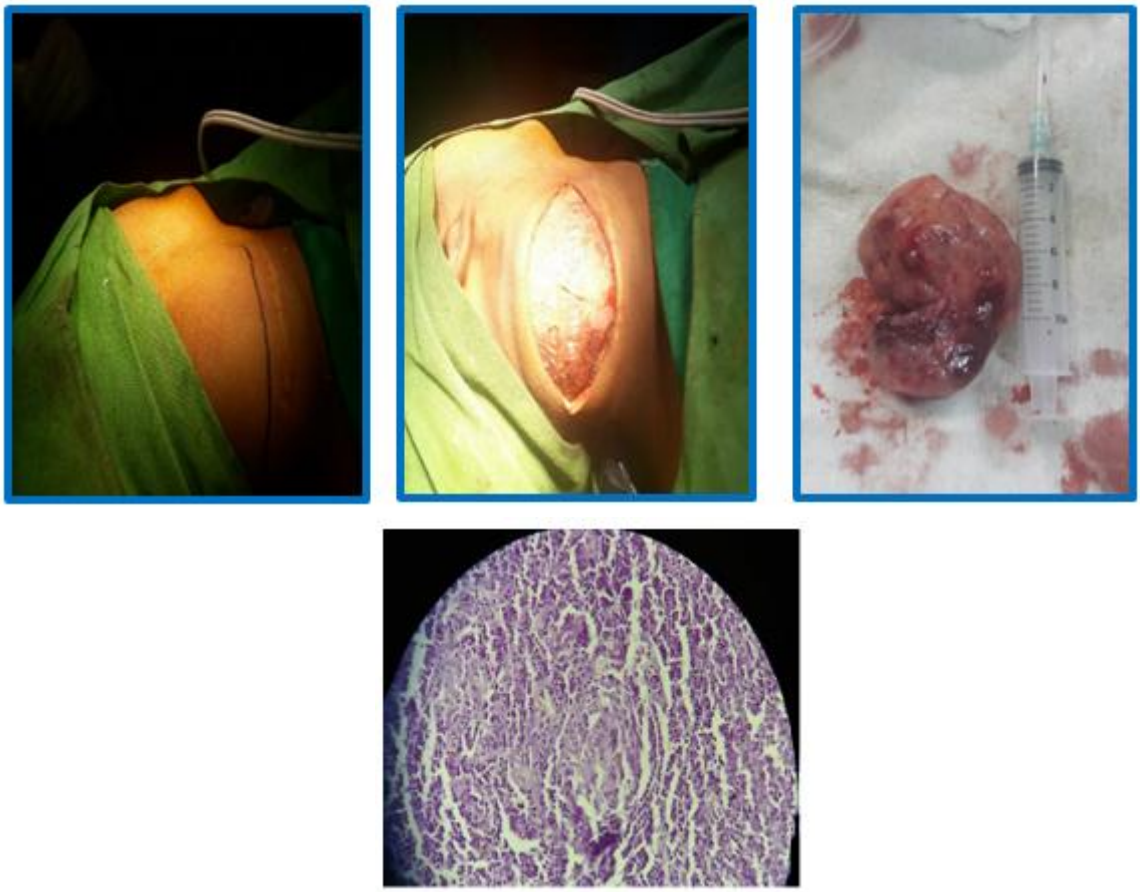
A 33yrs old female presented with a history of progressively enlarging mass in her right sided posterior triangle cervical region for last 6 months. She was largely asymptomatic except for an occasional discomfort around the mass. She had undergone an excisional biopsy from the same region 4 months back, in a different institute, the histopathological report then showed features suggestive of chronic granulomatous inflammation which was suggestive of tuberculosis. Based on this report she had been taking anti-tubercular drugs but the mass showed no signs of remission and was progressively enlarging.

On local examination a single non-tender, 10x5x3cm, firm mass was palpated which was slightly mobile and free from overlying skin. It was non tender and there was no local rise of temperature. ENT examination and general examination showed no abnormality.

All her blood investigations were within normal limits and viral serology for HIV, Hepatitis B and C were non-reactive.
A CT scan was done which showed a 11x5x2.5cm well circumscribed mass along the superior and lateral margin of sternocleidomastoid.



A surgical excision of the mass was carried out under general anaesthesia and histopathology showed hyaline-vascular variant of Castleman's disease.



Histopathology showed lymph nodal tissue with distorted follicles composed of atretic germinal centres and thickened mantle zone in an onion skin pattern.Hyalinisation of blood vessels noted.

CASE NO. 2

A 30yrs old female presented with a history of progressively enlarging mass in her right sided submandibular region for last 4 months. She too was largely asymptomatic except for an occasional discomfort around the mass.

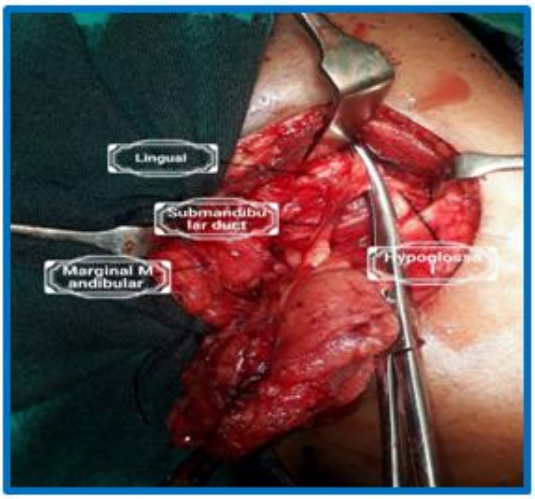
She had undergone a FNAC from the mass and the report then showed features suggestive of non-specific lymphadenitis. Based on this report she was given empirical antibiotic therapy, but the mass showed no signs of remission and was progressively enlarging.

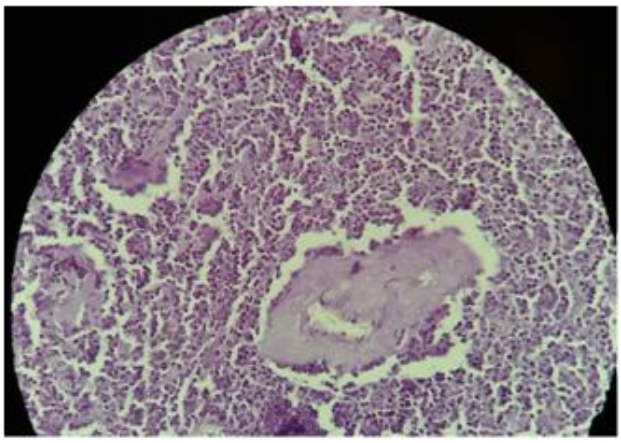
On local examination a single non-tender, 3x4cm, firm mass was palpated which was slightly mobile. All her blood investigations were normal. It was non tender and there was no local rise of temperature. ENT examination and general examination showed no abnormality.

All her blood investigations were within normal limits and viral serology for HIV, Hepatitis B and C were non-reactive.

An ultrasonography was done which revealed a single enlarged (5x4cm) submandibular lymph node with almost normal submandibular salivary gland, also no sailolith was detected.

A surgical excision of the mass was carried out under general anaesthetic. During the excision it was found that the node was firmly adherent to the submandibular gland hence a decision was made to decide the node along with the gland.





The histopathology report later showed hyaline-vascular variant of Castleman's disease.

III. Discussion

Castleman disease is a rare clinicopathological disease characterized by hyperplasia of lymph nodes and capillary proliferation [3]. It usually affects adolescents and young adults [3]. Both genders are equally affected [4].

The exact aetiology is unknown, but the current concepts are that it is an inflammatory response to an antigenic stimulus with increased production of IL6 and multicentric Castleman disease in HIV infected individuals is caused by HHV8 infection [5].

There are two clinical types of this disease, one is localized and usually benign, the other is multicentric and has a worse prognosis and may lead to the development of non-Hodgkin's lymphoma. Histologically it has been classified as hyaline vascular type, plasma cell type and lastly mixed type [3].

Clinically in localized type there is involvement of a single lymph node station. Most commonly it presents as a mediastinal mass (alternate sites include intra-abdominal masses or involvement of cervical, axillary, and inguinal nodes) [1]. The patients are usually asymptomatic unless a compression on neurovascular site or other organs cause symptoms.

In multicentric type multiple lymph node stations are affected with generalized lymphadenopathy. Associated symptoms may include fever, night sweats, fatigue, anorexia, and weight loss [3].Hepatomegaly and/or splenomegaly occur in 90% of patients [6].

Treatment in cases oflocalized type is surgical excision which is usually therapeutic. In cases of Idiopathic Multicentric type anti-IL-6 medications with steroids are used. (7). In HHV8 associated multicentric variety treatment with Ganciclovir or the anti-CD20 B cell monoclonal antibody may improve the outcome in some patients [6].

IV. Conclusion

Any atypical head and neck mass not responding to conservative management must be subjected to early excisional biopsy for definitive diagnosis, which can also be therapeutic at times. And no matter how rare the disease each and every clinical and pathological differential diagnosis must be kept at the back of our head during diagnosis of any head and neck mass so that no stone is left unturned , thereby helping the patient with early diagnosis and treatment.

References

[1]. Frizzera G, Massarelli G, Banks PM, Rosai J. A systemic lymphoproliferative disorder with morphological features of Castleman's Disease; Pathological findings in 15 patients. Am J Surg Pathol 1983; 7: 212–31.
[2]. Beraldo S et al. Castleman disease as an uncommon cause of a neck mass. Acta Laryngologia 2006;126:108-11.
[3]. Watkinson J, Gilbert RW, editors. Stell and Maran's text book of head and neck surgery and oncology. 5th ed. London: Hodder Arnold; 2012, Chapter 12, p. 230.
[4]. Simpson D. Epidemiology of Castleman disease. Hematol Oncol Clin North Am. 2018;32(1):1-10.
[5]. Wu D, Lim MS, Jaffe ES. Pathology of Castleman disease. Hematol Oncol Clin North Am. 2018;32(1):37-52.
[6]. Dispenzieri A, Gertz MA. Treatment of Castleman's disease. Curr Treat Options Oncol. 2005;6:255–266.
[7]. Dispenzieri A, Gertz MA. Treatment of Castleman's disease. *Curr Treat Options Oncol*. 2005;6:255–266.

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