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Castleman Disease: An Unheard Entity in the Neck

Priti S Hajare[1], Jahnavi[2], Suhasini[3], Adarsh Sanikop[4]

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Author Affiliation: [1]Professor, [2,3]Postgraduate, Department of ENT and HNS, [4]Assistant professor, Department of Pathology, Jawaharlal Nehru Medical College, K.L.E. University Campus, Nehru Nagar, Belgaum, Karnataka-590010, India.

Corresponding Author: Jahnavi, Postgraduate, Department of ENT and HNS, Jawaharlal Nehru Medical College, K.L.E. University Campus, Nehru Nagar, Belgaum, Karnataka-590010, India.

E-mail: jahnavi.bp79@gmail.com

Abstract

Castleman Disease (CD) is a nonclonal lymphoproliferative disorder that can affect either a single lymph node called unicentric or, can be generalized, multicentric. Though the disease has a wide spectrum of presentation, still there are no studies describing its natural history. Unicentric type is usually a chronic and asymptomatic swelling with neck being the unusual site as compared to multicentric type which presents with mediastinal lymph node involvement and constitutional symptoms. Here we report a 24 year male, presenting with long standing history of painless solitary gradually progressive swelling in the posterior triangle of the neck, which was treated successively with excisional biopsy and was diagnozed with histopathology which showed unicentric type of Castleman disease. He was evaluated further to rule out multicentric type of disease, no other swellings seen in the body.

Keyword: Castleman disease; Excisional biopsy; Unicentric.

Introduction

Castleman Disease (CD) is a rare, poorly understood lymphoproliferative disease. It is otherwise known as "giant lymph node hyperplasia", "lymph node hamartoma", "angiofollicular mediastinal lymph node hyperplasia", and "angiomatous lymphoid hyperplasia"[1]. It is a rare disease of unknown aetiology that affects lymphatic tissues in diverse locations.[2] Castleman disease is classified into unicentric and multicentric type clinically, based on the number of lymph nodes involvement and histologically further classified into hyaline vascular variant, plasma cell variant, mixed cellular or plasmablastic variant. Unicentric type is usually chronic and asymptomatic whereas multicentric type usually presents with constitutional symptoms. The definitive treatment is based on pathological findings postoperatively. The gold standard management is by complete surgical excision. The main aim of our study is to describe a case of unicentric type of Castleman's disease, its presentation, investigations, and the intraoperative management.

Case Presentation

A 24 year old male presented with a 5x4 cm ovoid, painless, gradually progressive swelling in the left

posterior triangle of the neck since 10 years, Fig. 1 with well - defined borders, which was non tender, firm in consistency and mobile in both horizontal and vertical plane, the plane of the swelling was above the sternocleidomastoid muscle. The skin over the swelling was normal and pinchable with no other associated symptoms. No other swellings palpated elsewhere.

Fig. 1: Clinical photograph

He was subjected to routine investigations, *ultrasound neck* and *fine needle aspiration cytology* of the swelling which was *inconclusive*, he then underwent excisional biopsy of the swelling and it was sent for histopathological examination.

Fig. 1: Clinical photograph – Showing the swelling in the left lateral aspect of the neck

Management

Under General Anesthesia, a horizontal skin incision around 5 cm was taken over the swelling in the posterior triangle of the neck. Skin, superficial fascia and sternocleidomastoid muscle was separated, capsule over the mass identified and delineated completely all around and the mass was separated from all the adjacent structures and from the base which was anterior border of the trapezius muscle and removed en-mass Fig.2 A and B. He was followed up for one year and he was asymptomatic.

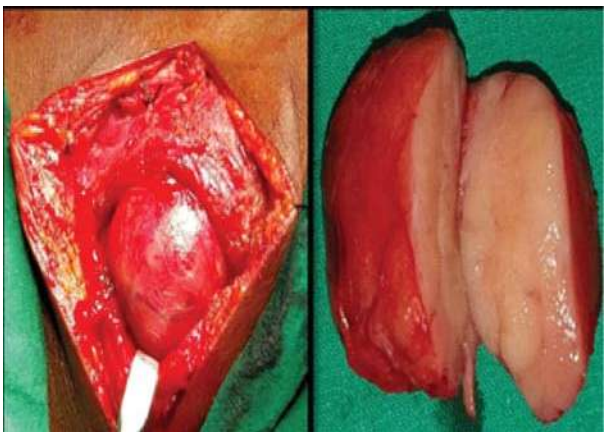

Fig. 2: Globular soft tissue mass measuring 5x4.5x3 cm, external surface capsulated, Cut section – homogenous, greyish- white in color.

B - Cut section

Histopathology

10x- shows large or normal-sized lymphoid follicles with germinalcentres.It has conserved polarity and paracortical vascular proliferation with some vessels of the paracortex penetrating radially towards the germinal centers, forming lesions that resemble a spiral palette (lollipop appearance). [Fig 3A]

100x-shows magnified image of lymphoid follicles with two or more germinal centers referred to as "twin germinal centers" (twinning). The germinal center has central fibrosis and small lymphocytes with concentric ring formation giving an "onion skin"appearance. [Fig 3B]

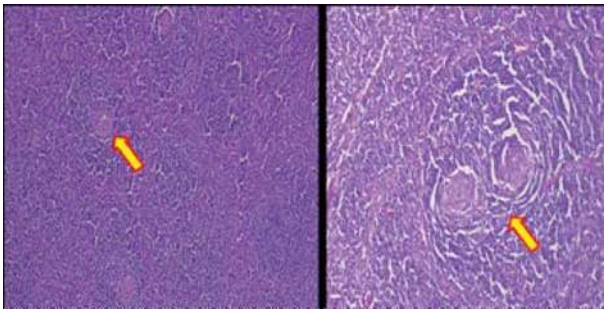

Fig. 3: a – 10 X – Lollipop appearance
3b- 100 X- Twinning and onion skin appearance

Discussion

In 1954, Benjamin Castleman first reported, on 2 patients with isolated mediastinal lymph node involvement with lymphoid follicles showing germinal-center involution and increased capillary proliferation with hyperplasia of the endothelium in both follicular and interfollicular regions[1]. The etiopathogenesis is unknown, but based on few studies, evidence points toward faulty immune regulation, resulting in increased B-lymphocyte and plasma-cell proliferation in lymphoid tissue. 1-3 Interleukin [6] (IL-6) plays a pivotal role [2,3].Mediastinum is the most common site of presentation,[2,4,5] Other sites include the neck, axilla, mesentery, pelvis, pancreas, adrenal gland, and retroperitoneum. [2,4,5] The disease in most cases has aninflammatory background, with predominent levels of interleukin-[6] (IL-6) and vascular endothelial growth factor (VEGF).[6,7]

The lymph nodes with small, prominent, hyalinized follicles with marked interfollicular vascular proliferation are categorized into hyaline vascular type and is further classified into Unicentric type of Castleman's Disease (UCD).[8]

Multicentric Castleman's Disease (MCD) has a systemic involvement with symptoms characterized by fever and night sweats with generalized peripheral lymphadenopathy and hepatosplenomegaly, and is usually a plasma cell variant.[9]

Although the incidence of the disease is unknown, but because of its association with Human Immunodeficiency Virus (HIV) and Human Herpes Virus (HHV-8), MCD is also seen to be associated with few malignancies, particularly lymphomas and Kaposi's sarcoma. It is seen that nowadays Multicentric type of disease is resulting in fatality in few patients due to disease progression, disseminated infection, or related malignancies.[10]

A wide variety of treatment modalities have been tried for MCD along with surgery, steroids, specific antibodies, antiviral agents, radiation, cytokine inhibitors, and even chemotherapy. Usually, in these conditions, surgery has no role in the treatment of MCD, although splenectomy has shown symptomatic improvement temporarily in few patients. Unicentric type of disease is the commonest type among the two and is characterized by a localised benign lymphoproliferative disorder in young adults and is not associated with Human Herpes virus-8 infection and usually managed with surgical resection.[10]

Conclusion

Achieving the diagnosis is quite complicated as there is no proper laboratory investigations or imaging modalities specific for this disease except for histopathologic examination.The gold standard mode of diagnosis is excisional biopsy from an affected lymph node.[8,9] as done in our case. The treatment mainly depends on the type of Castleman disease. Unicentric type has a good outcome to excisional biopsy.[9] Radiotherapy can also be considered with successful outcomes in patients with either residual disease after incomplete resection or unresectable lesions. Chemotherapy is usually not indicated for unicentric type of disease. To conclude prognosis of unicentric Castleman's disease is excellent.

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