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### Castleman's Disease: A Case Report and Brief Review of the Literature

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### Short Communication

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#### ABSTRACT

Castle man's diseases (CD), also called as giant lymph nodal hyperplasia is a relatively rare condition of uncertain etiology It involves massive proliferation of lymphoid tissues, and may present as nodal or extra nodal mass which may be confused with other causes of lymphadenopathy on clinical as well as gross examination. This diseases mimic's lymphomas and TB. We report a case of multicentric CD, in a patient who was initially suspected to have disseminated kochs but was later histologically confirmed to have Castleman's disease. In this report we also attempt to provide insight through the review of medical literature into the clinical features, pathogenesis and management of this disorder.

#### INTRODUCTION

CD also called as angiofollicular hyperplasia is a rare benign systemic lymphoproliferative disorder characterised by a peculiar form of lymphnode hyperplasia [1,2,3]. It was first described by Benjamin Castleman in 1956. CD can be classified as a) unicentric Vs multicentric based on clinical and radiological features b) hyaline vascular Vs plasmacytic mixed cellularity type based on histopathology. It can be found wherever lymphnodes are present but commonly involves mediastinum or lung hilum but can occur in other spaces like pelvis, neck, retroperitoneum and muscle [4]. The etiology is unknown, the two main hypotheses being abnormal immune response and viral infection. CD affects patients of varying ages and cases have been reported from adolescence to seventies with a clear male preponderance [5]. Most cases of CD represent either the hyaline vascular variant (80-90%) or plasma cell variant (10-20%) and a small percentage of patients with mixed histologic appearance. Patients with HVP may exhibit no symptoms or only lymphadenopathy while plasma cell type disease presents with fever, weightloss, rash, anaemia, hepatosplenomegaly and night sweats [6]. Treatment varies from curative surgery for HVP type of CD to the use of chemotherapy for multicentric CD.

#### CASE REPORT

A 52 Year old immuno competent, male presented with fever, low to moderate grade, intermittently for 2 yrs with generalised LAP, he also complained of malaise, decreased appetite and wt loss (5-6 kg in 2 yrs).

He was treated as a case of disseminated kochs by 4 drug regime ATT for 6 months without much relief. Physical examination revealed an average built male with normal BP and pulse rate, having bilateral cervical, axillary and inguinal lymphadenopathy with splenomegaly. All routine lab tests like CBP, blood urea, serum creatinine, hepatic functions were within normal range. Chest XRay showed paratracheal and bilateral hilar lymphadenopathy (fig1). Ultrasound of abdomen showed enlarged spleen with retroperitoneal LAP. A CT scan of thorax and abdomen showed few subcentric lymph nodes in right paratracheal region, mediastinum and retro peritoneum with no evidence of calcification or necrosis within these nodes. Fine needle aspiration cytology of inguinal lymph nodes showed atypical lymphoid proliferation. An excisional biopsy of axillary LN showed typical picture of hyalinised follicles surrounded by lymphocytes arranged in "onion skin" layer like pattern in lymphnode, leading to conclusion of Caselman's disease( hyaline vascular type) (Fig 2). The patient was referred to a hematologist and was put on steroid therapy which successfully induced disease remission and brought symptomatic relief.

Fig1: Chest x-ray PA view shows Paratracheal and bilateral hilar LAP

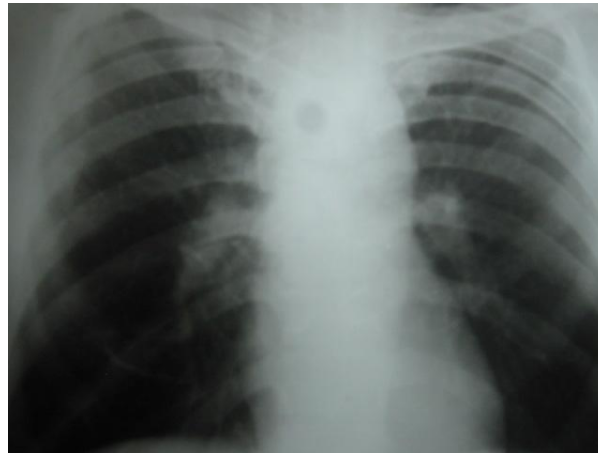
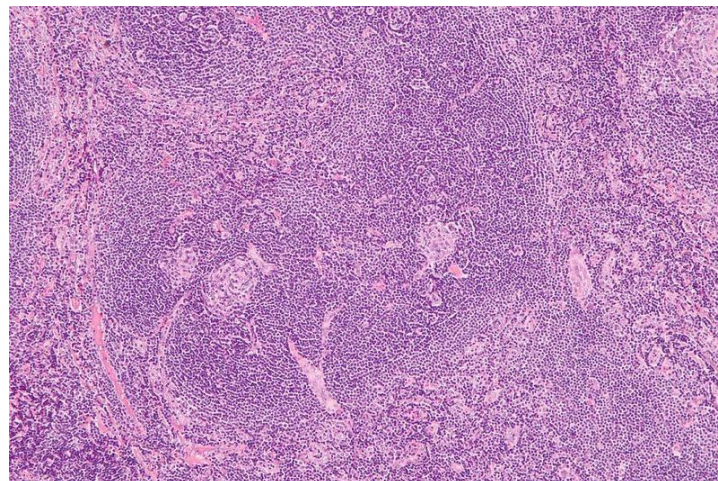


Fig 2: Slide shows lymphoid follicles scattered throughout node with abnormal germinal centres and marked vascular proliferation and hyalinisation arranged in onion skin layer like pattern suggestive of hyaline vascular type CD



## DISCUSSION

Castleman's disease (CD) is a lymphoproliferative disorder which is histologically characterised by angiofollicular lymph node hypertrophy<sup>[7]</sup>. Our case report along with a review of existing literature attempts to provide new insight into this rare and benign disorder with overlapping clinical features and histological variants. CD has three histological variants (hyaline vascular, plasma cell and mixed type) and two clinical types (localised and multicentric). The localised hyaline vascular type is seen in 90% cases of localised disease. The etiology is unknown, the two main hypotheses being abnormal immune response and viral infection<sup>[8]</sup>. Frizzera et al studied clinical and histopathological findings in 15 patients of CD and opined that despite some similarities with autoimmune diseases, the main features of CD are a hyperplastic-dysplastic lymphoid disorder in a setting of immunoregulatory deficit<sup>[9]</sup>. The differential diagnosis mostly includes all reactive hyperplasias of lymph nodes, Hodgkin's lymphoma. Hyaline vascular CD is difficult to diagnose on FNAC and may be mistaken for lymphoreticular malignancy because of presence of cells having nuclei showing atypical features<sup>[10]</sup>. The prognosis of localised CD is good with surgery. The multicentric CD, as in our patient is a systemic disease with disseminated lymphadenopathy, hepatosplenomegaly and constitutional symptoms. The course of multicentric CD is unpredictable, it

can be stable or can be characterised by remissions and exacerbations and can be fatal some times, because of infections and malignancies (lymphoma and Kaposi's sarcoma). HHV-8 also called as Kaposi's sarcoma associated herpes virus is linked to multicentric CD and Kaposi's sarcoma especially in patients with HIV. In conclusion the cytogenetic changes in stromal cells of the hyaline vascular type and the role of IL-6 and HHV-8 in the pathogenesis of the plasma cell type suggest that both are separate diseases. Surgical removal of affected lymph node is the treatment of choice, if it is not possible as in multicentric CD then radiotherapy is beneficial. In the plasma cell and multicentric type corticosteroids, chemotherapeutic drugs, monoclonal anti bodies and immunomodulators may be used<sup>(11)</sup>. long term follow-up of patients with CD shows low or nil recurrence in successfully treated cases .

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