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CASE STUDY

MULTICENTRIC TYPE CASTLEMAN'S DISEASE – A CASE REPORT

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ABSTRACT

Introduction: Castleman's disease is a rare lymphoproliferative disorder of unknown etiology with clinical manifestations ranging from asymptomatic discrete lymphadenopathy to recurrent episodes of diffuse lymphadenopathy with severe systemic symptoms.
Case report: We report a case of multicentric Castleman's disease presenting as anasarca. The patient had predominant systemic involvement during the clinical course. Patient was started on chemotherapy(CHOP regimen). Patient succumbed to sepsis three months after diagnosis. This case has been reported for its rarity.
Conclusion: Though Castleman's disease is a relatively rare entity, its varied clinical manifestations especially multicentric Castleman's disease may cause diagnostic confusion.

INTRODUCTION

A 39yr old male patient presented with 6 months history of progressive anasarca, breathlessness, loss of appetite and excessive sweating (Figure 1). Examination revealed anasarca, generalised lymphadenopathy and moderate hepatosplenomegaly, polyneuropathy. Evaluations directed at chronic liver disease, chronic kidney disease, congestive cardiac failure, autoimmune diseases and Tuberculosis were negative.

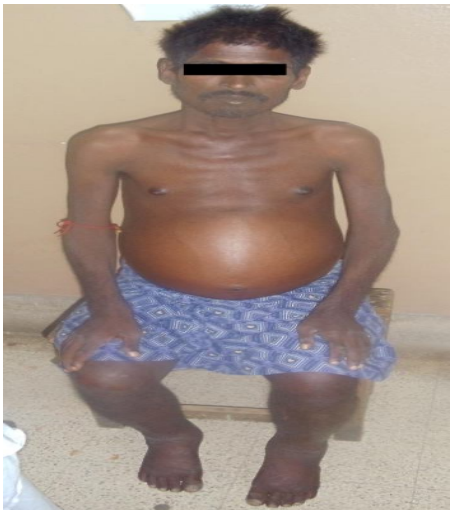

Figure 1. Patient with castleman disease

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Left axillary lymph node biopsy revealed reactive follicles along with follicular cells covered with concentric layers of lymphocytes with perivascular hyalinization (Figure 2).

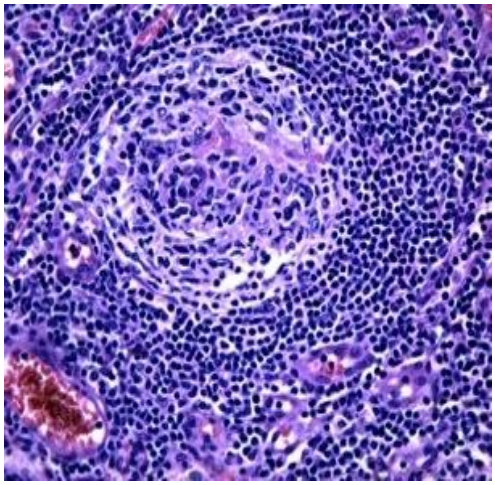

Figure 2. Reactive follicle covered by concentric layer of lymphocytes with perivascular hyalinisation

There were also interfollicular plasma cells (Figure 3). It was classified as mixed type multicentric castleman disease based on histology and immunohistochemistry (Figure 4). Bone marrow biopsy showed plasmacytosis (15%). CT Abdomen & Pelvis showed multiple enlarged lymph nodes and multiple osteoblastic lesions in spine, pelvic bones, ribs and gross ascitis (Figure 5).

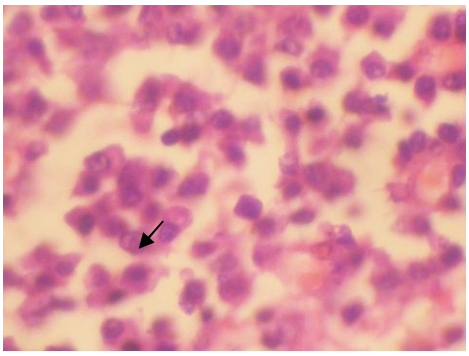

Figure 3. Abundant interfollicular plasma cells

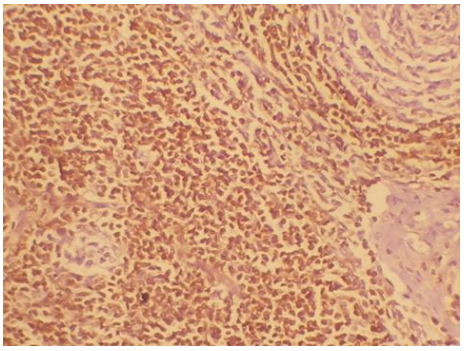

Figure 4. Bcl-2 negative reactive germinal centre

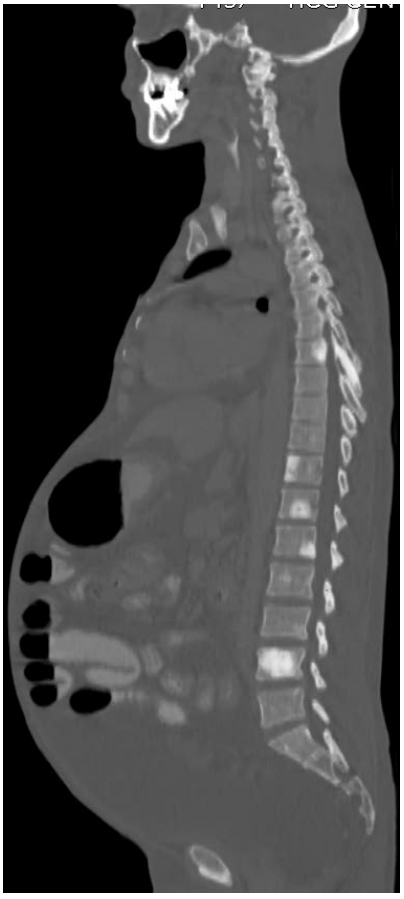

Figure 5. CT scan showing osteoblastic vertebral metastasis

Whole body PET SCAN showed multiple enlarged metabolically active lower cervical, axillary, mediastinal, bilateral iliac, inguinal and mesenteric lymph nodes and extensive osteoslerotic skeletal lesions with no obvious primary lesion (Figure 6).

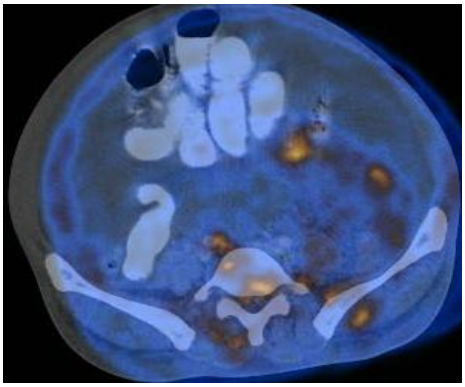

Figure 6. PET SCAN showing increased FDG concentration in enlarged bilateral iliac lymph nodes

ELISA for HIV was negative. Serum electrophoresis showed polyclonal gammopathy and was negative for M spike. Patient was put on CHOP regimen chemotherapy. Reassessment with PET scan after two months showed significant improvement in the form of decrease in metabolic activity of lymph node burden. Patient succumbed to sepsis three months after diagnosis.

DISCUSSION

Castleman's disease was originally described by Dr. Benjamin Castleman in 1954 (Castleman and Towne 1954). The etiopathogenesis of this disease is not completely understood. Current knowledge about the disease suggests that dysregulation of IL-6 mediated pathways leads to abnormal non-clonal lymphocyte proliferation. Castleman's disease is classified as hyaline vascular variant, plasma cell variant or mixed cellularity variant (Keller *et al*., 1972). Castleman's disease can also be classified based on clinical presentation as localized or unicentric type and systemic or multicentric type of Castleman's disease which is less common and mostly of the plasma cell variant. Our case is a multicentric type Castleman's disease which is of mixed cellularity variant. Patients with multicentric Castleman's disease often require systemic treatment and their prognosis is less favourable than those for unicentric Castleman's disease (Casper 2005). Currently no standard treatment exists for Castleman's disease. New molecular targeted therapies may provide solution in the future.

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