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Castleman's disease: A case report

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Abstract : Castleman's disease (CD) is rare lymphoproliferative disorder characterised histologically by non-neoplastic angiofollicular lymph node hypertrophy .CD has two distinct diseases (unicentric and multicentric) with very different prognoses. It usually associated with malignancies including Kaposi sarcoma, lymphoma, and POEMS syndrome. Viral infection may also be associated such as HIV/HHV-8 infection. MCD is a systemic disease characterized by fever and night sweats associated with generalized peripheral lymphadenopathy and hepatosplenomegaly, which is frequently related to the plasma cell variant. A Case report of a 55-year-old Saudi female presented with hematemesis. Her past medical history revealed that she was suffering from undiagnosed generalized lymphadenopathy with hepatosplenomegaly for 30 years accompanied by intermittent episodes of fever. She tested positive for CMV. Lymph node biopsy showed reactive lymphoid hyperplasia with Castleman's like feature. This case report aims to provide the importance of obtaining a definitive histological diagnosis in patients with generalized lympadenopathy associated with systemic manifestations. Furthermore, to be aware of the possible long-term complications that MCD Patients might present with.

Keywords: Castleman's disease, angiofollicular lymph node hypertrophy, lymphadenopathy , hepatosplenomegaly

Background:

Castleman's disease is a sporadic lymphoproliferative disorder characterized histologically by non-neoplastic angiofollicular lymph node hypertrophy. It has two distinct types; unicentric and multicentric with very different prognoses. It usually associated with malignancies including Kaposi sarcoma, lymphoma, polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes (POEMS) syndrome and viral infections such as Human immunodeficiency virus/Human herpes virus (HIV/HHV-8) infection. Metacentric Castleman's disease is a systemic disease characterized by fever and night sweats associated with generalized lymphadenopathy and hepatosplenomegaly, which is linked to plasma cell variant.[1]

Case presentation:

A 55-year-old female came to the emergency department complaining of hematemesis. She had a history of abdominal distension and shortness of breath for thirty years and had been treated as inflammatory bowel syndrome (IBS). Also, she had generalized lymphadenopathy with hepatosplenomegaly accompanied by intermittent fever episodes for which she had a medical consultation however they were unable to establish a diagnosis.

On examination pulse 118/min, BP 90/60 mmHg, afebrile, evident pallor,

drumstick fingers, and generalized lymph nodes enlargement but no jaundice. Chest: Basal decrease air entry, S1+S2+gallop with a mid-systolic murmur. Abdomen: Liver span 17 cm with 15 cm splenomegaly and massive ascites.

Laboratory investigation showed WBC 4.23 10^3/ml with low lymphocyte and basophils absolute count. Albumin 28 g/L, creatinine 30 umol/L. Direct bilirubin 19 umol/l and total bilirubin 34.3 umol/l. GGT 175 U/L. Rheumatoid factor 9.75, Hepatitis work up, HIV-Ab, HSV-Ab, HHV8, EBV (IgG and IgM), mono-spot test, TB Skin test were all negative, IgM 0.944, IgA 4.43, IgG 14.8. Antiphospholipid IgG Positive (15.5) while Antiphospholipid IgM was negative. CT-scan of Chest, Abdomen, and Pelvic showed Extensive mediastinal and abdominal lymphadenopathy, Hepatosplenomegaly and splenic varices with massive ascites. Moreover, Multiple hepatic focal lesion and bilateral basal lung consolidation with pleural effusion were also seen. Histopathology of the axillary LN Biopsy showed Reactive lymphoid hyperplasia with Castleman disease like feature.

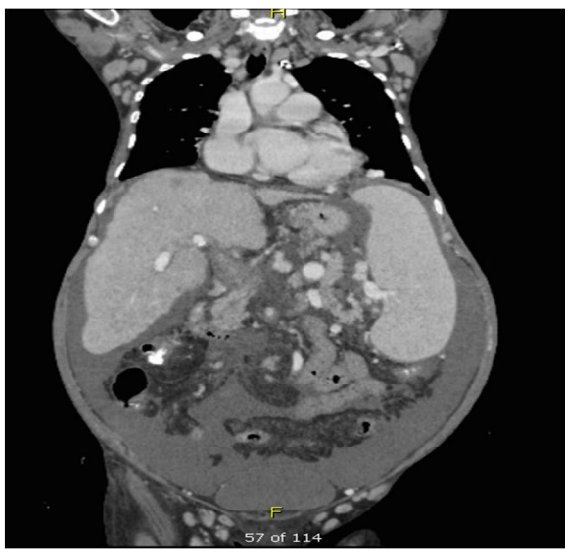

Figure1: coronal section CT showing hepatosplenomegaly

Discussion:

Castleman's disease is a rare clinicopathological lymphoproliferative chronic disorder characterized histologically by non-neoplastic angiofollicular lymph node hypertrophy and hyperplasia.[2] [3] [4] [5] Benjamin Castleman first described it in 1956.[6] The incidence is unknown. It has been reported mostly in adults however it can appear at any age.[3]

Although the etiology remains obscure, however, there are several theories which explain the etiology of the disease. Castleman's disease could be associated

with HHV-8 infections, Kaposi's sarcoma and HIV infection [3 5].

Clinically Castleman's disease has two types unicentric Castleman disease and multicentric Castleman's disease. Unicentric Castleman's disease is more common than multicentric Castleman disease and presents early in adulthood. Additionally, it rarely causes any systemic symptoms. However, multicentric Castleman's disease involves multiple sites and found in the 5th and 6th decade of life. Multicentric Castleman's disease usually presents with vague constitutional symptoms includes fever, fatigue, night sweat and weight loss [5]. Histologically there are three types: Hyaline vascular, Plasma cell variant and mixed of both previous types. The hyaline vascular is usually found in unicentric Castleman's disease while plasma cell variant and mixed types are mostly associated with multicentric Castleman's disease. [3]

The unicentric Castleman's disease is usually asymptomatic and discovers accidentally in routine chest X-ray or presents with discomfort due to local compression. The common sites are the abdomen, peripheral lymph node, and mediastinum. Multicentric Castleman's disease presents with multiple lymph node hyperplasia and systemic symptoms that are linked to elevated IL-6 production.[3] Histological study of the enlarged lymph node is required for both types to confirm the diagnosis. [7]

Unicentric Castleman's disease commonly has a good prognosis and treated with simple local excision of the affected lymph node while multicentric Castleman's disease has different methods of managements including surgical removal, chemotherapy, and steroids.[7]

Conclusion:

This case report aims to provide the importance of obtaining a definitive histological diagnosis in patients with generalized lymphadenopathy associated with systemic manifestations. Furthermore, to be aware of the possible long-term complications that multi-central Castleman's disease Patients might have.

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