

PICTORIAL

GROSS HEPATOSPLENOMEGALY IN A PATIENT WITH HUMAN HERPES VIRUS-8 ASSOCIATED MULTICENTRIC CASTLEMAN'S DISEASE

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Figure-1: CT scan demonstrating gross hepatosplenomegaly

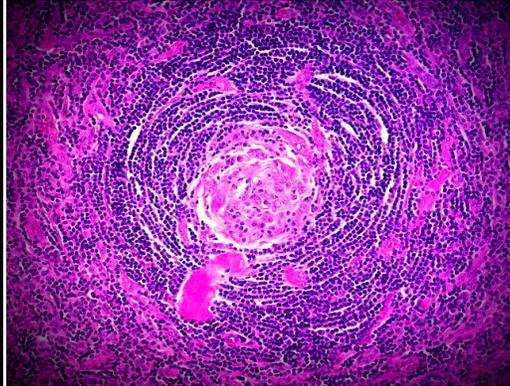


Figure-2: Histopathology of the lesion

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A 30 year old man presented with a three week history of worsening dyspnoea, productive cough and fever. Examination revealed cervical lymphadenopathy, splenomegaly and bilateral cutaneous lesions on the legs. The patient denied any medical co-morbidities or any recent travel. Blood tests showed anaemia (haemoglobin of 76 g/L), thrombocytopenia ($45 \times 10^3/\text{mm}^3$) a white blood cell count of $5.1 \times 10^3/\text{mm}^3$ and a CRP of 61 mg/dL. A HIV test was positive with a CD4 count of 12 count cells/ mm^3 and a HIV Viral Load of 1420045 copies/ml.

A punch biopsy was taken from the leg lesions was positive for HHV-8 in keeping with the diagnosis of Kaposi Sarcoma. Computed tomography (CT) scan of the thorax and abdomen (Figure-1) demonstrated mediastinal lymphadenopathy, lungs nodularity and bilateral patchy opacities, and bronchoscopy revealed Kaposi sarcoma lesions in the trachea and main lobar bronchi. A cervical lymph node showed marked plasmacytic hyperplasia with focal EBV positivity and regression of germinal centers, in keeping with a diagnosis of HHV-8-associated multicentric Castleman's disease. Investigations for atypical pneumonia, TB and PCP were all negative.

Despite the patient receiving antiretrovirals, antibiotics, steroids and blood transfusions the patient developed multi-organ failure and died shortly after.

Multi-centric Castleman's disease (MCD) is rare lymphoproliferative disorder, with 50% of these cases caused by HHV-8, which also causes Kaposi Sarcoma.¹ It presents with generalized lymphadenopathy, hepatosplenomegaly, anaemia, hyper-immunoglobulinaemia, and systemic symptoms such as fevers, night sweats and weight loss.²

Diagnosis of Castleman's disease is based on lymph node biopsy which can demonstrate Hyaline-Vascular, plasmablastic and plasma Cell variants of Castleman's disease. (Figure-2)

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