iMCD/TAFRO SYNDROME- A CASE REPORT

Castleman's disease (CD) was first described in a group of patients with benign localised hyperplastic lymph-nodes in 1956 by Castleman et al. The 3 subtypes of CD are Unicentric CD (UCD), Human herpesvirus 8 (HHV-8)–associated multicentric Castleman disease (HHV-8-associated MCD) and HHV-8–negative/idiopathic multicentric Castleman disease (iMCD). TAFRO syndrome is a clinicopathologic variant of iMCD characterized by Thrombocytopenia, Ascites (Anasarca), myeloFibrosis, Renal dysfunction, and Organomegaly. We report a case of TAFRO syndrome in an HIV-negative 20 year old black male who presented with generalised lymphadenopathy, fever, normocytic anemia, thrombocytopenia, and acute renal insufficiency. History of being treated as sickle cell disease since the age of 16 years. CRP was markedly elevated and had marked hypergammaglobulinemia. Hb electrophoresis was normal. Chest and abdominal CT revealed multiple air filled cysts in the lungs, hepatosplenomegaly, and multiple mildly enlarged lymph nodes. An excisional biopsy of inguinal lymph node showed onion lollipop lesions and reactive follicular hyperplasia with onion skin architecture in keeping with Castleman's disease-mixed plasmacytosis and hyaline . HHV-8 was negative. Bone marrow biopsy showed megalobalsts and less than 5% plasma cells. This case report show the need to raise awareness on CD

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