“PEELING BACK THE ONION LAYERS” - THE CHALLENGE OF HIV ASSOCIATED MULTICENTRIC CASTLEMAN’S DISEASE.

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Case presentation:
A 66 year old Japanese-born man, self-identified MSM, with virologically suppressed HIV on a stable antiretroviral regimen of abacavir/lamivudine/nevirapine for 10 years, presented with biopsy-confirmed severe Stevens Johnson Syndrome/Toxic Epidermal Necrolysis requiring management on the Burns ward. The only identifiable precipitant was aspirin taken 2 weeks prior for mild headache. Post discharge, he experienced fevers, lethargy and loss of weight and developed widespread lymphadenopathy with new erythematous papular rash on bilateral upper limbs 8 weeks after discharge. Investigations revealed pancytopenia (significant neutropenia (0.06 x10^9/L) and raised inflammatory markers. Parvovirus B19, CMV and EBV serology were indicative of past infection, autoimmune screen, LDH, beta-2-microglobulin and free light chain ratio were normal. Bone marrow aspirate/trephine displayed a reactive hypercellular aspirate without evidence of lymphoproliferative disorder.

Due to ongoing symptoms without diagnosis, a Positron Emission Tomography scan revealed extensive widespread FDG-avid lymphadenopathy. Differentials included T cell lymphoma, haemophagocytosis lymphohistiocytosis (HLH) or Castleman’s Disease. A core biopsy of an axillary lymph node revealed an ill-defined onion-skin pattern of lymphocytes on histopathology, with scattered plasma cells immunoreactive to Human Herpes Virus-8 (HHV-8) immunoperioxidase stain. Concurrently, HHV-8 was detected in blood by polymerase chain reaction, confirming a diagnosis of HIV-associated Multicentric Castleman’s Disease (MCD).

He was finally treated with rituximab once weekly for 4 weeks, in combination with therapeutic-dose valganciclovir. He has experienced improvement in all cell lineages, and constitutional symptoms have also abated.

HIV-associated MCD is rare, with an apparent increased incidence in the ART era. The manifestations, diagnosis and pathogenesis, including the relationship to HHV-8 and association with pro-inflammatory cytokines, specifically interleukin-6 will be discussed. It has a poor prognosis if left untreated. Guidance for treatment is limited by small clinical trials. Tociluzimab and rituximab appear promising, as does anti-viral therapy (ganciclovir or valganciclovir).

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