World Academy of Science, Engineering and Technology
International Journal of Pharmacological and Pharmaceutical Sciences
Vol:9, No:12, 2015

Bulbar Conjunctival Kaposi's Sarcoma Unmasked by Immune Reconstitution Syndrome

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Abstract : Kaposi's sarcoma (KS) is the most common HIV-related cancer, and ocular manifestations constitute at least 25% of all KS cases. However, ocular presentations often occur in the context of systemic KS, and isolated lesions are rare. We report a unique case of ocular KS masquerading as subconjunctival haemorrhage, and only developing systemic manifestations after initiation of HIV treatment. Case: A 49-year old man with previous hypertensive stroke and newly diagnosed HIV infection presented with an acutely red left eye following repeated bouts of coughing. Given the convincing history of poorly controlled hypertension and cough, a diagnosis of subconjunctival haemorrhage was made. Over the next week, his ocular lesion began to improve and he subsequently started anti-retroviral therapy. Prior to receiving anti-retroviral therapy, his CD4+ lymphocyte count was 194 cells/mm3 with HIV viral load greater than 1 million/ml. This rapidly improved to a viral load of 150 copies/ml within 2 weeks of starting treatment. However, a few days after starting HIV treatment, his ocular lesion recurred. Ophthalmic examination was otherwise normal. He also developed widespread lymphadenopathy and multiple dark lesions on his torso. Histology and virology confirmed KS, systemically triggered by Immune Reconstitution Syndrome (KS-IRIS). The patient has since undergone chemotherapy successfully. Discussion: Kaposi's sarcoma is an atypical tumour caused by human herpesvirus 8 (HHV-8), also known as Kaposi’s sarcoma-associated herpesvirus (KSHV). In immunosuppressed patients, KSHV can also cause lymphoproliferative disorders such as primary effusion lymphoma and Castleman's disease (in our patient’s case, this was excluded through histological analysis of lymph nodes). KSHV is one of the seven currently known human oncoviruses, and its pathogenesis is poorly understood. Up to 13% of patients with HIV-related KS experience worsening of the disease after starting anti-retroviral treatment, due to a sudden increase in CD4 cell counts. Histology remains the diagnostic gold standard. Current British HIV Association (BHIVA) guidelines recommend treatment using anti-retroviral drugs, with either intralesional vinblastine for local disease or systemic chemotherapy for disseminated KS. Conclusion: This case is unique as ocular KS as initial presentation is rare and our patient's diagnosis was only made after systemic lesions were triggered by immune reconstitution. KS should be considered as an important differential diagnosis for red eyes in all patients at risk of acquiring HIV infection.

Keywords : human herpesvirus 8, human immunodeficiency virus, immune reconstitution syndrome, Kaposi’s sarcoma, Kaposi’s sarcoma-associated herpesvirus
Conference Title : ICMVM 2015 : International Conference on Molecular Virology and Microbiology
Conference Location : Bangkok, Thailand
Conference Dates : December 17-18, 2015