ORBITAL NK/T CELL LYMPHOMA AS FIRST PRESENTATION OF HIV IN OVERSEAS VISITOR

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Background: A 37 year old Zimbabwean female, visiting family in Australia, presented with facial swelling, fever and headaches. She was diagnosed with peri-orbital cellulitis, treated with intravenous antibiotics, single dose of dexamethasone and discharged on oral antibiotic therapy. She re-presented 6 days later with ongoing symptoms when a repeat commuted tomography scan demonstrated dehiscence of the medial wall of the orbit with a small subperiosteal collection at the lamina papyracea. Endoscopic sinus surgery drained the collection and biopsy result was consistent with an osteomyelitis. HIV Ag/Ab testing was performed due to delayed clinical improvement, risk factors and significant lymphopenia. HIV-1 antibody was confirmed by Western Blot with CD4 count 26 x10^6/L and viral load 281838 copies/ml. Treatment was with Atripla and prophylaxis for Pneumocystis jirovecii and Mycobacterium avium complex. Despite therapy, fevers persisted. Investigations revealed mild anaemia, lymphopenia and deranged liver function. Ferritin of >90,000µg/L raised suspicion of Haemophagocytic syndrome. A bone marrow biopsy demonstrated haemophagocytosis and repeat sinus biopsy diagnosed an extra-nodal NK/T cell lymphoma (CD2+/CD7+/CD56+ with Ki-67 80% and EBER+). Positron Emission Tomography confirmed stage 2 disease. She was treated with 1 cycle of Cyclophosphamide, Etoposide, Doxorubicin, Vincristine and Prednisolone (CHOEP) prior to returning to Zimbabwe.

Analysis: Haemophagocytic syndrome is a rare complication of HIV infection but has recognised association with lymphoproliferative disorders. HIV-associated lymphomas are typically aggressive B-cell disorders. The diagnosis of extranodal NK/T cell lymphoma of nasal subtype is rare in African populations. Epstein-Barr Virus (EBV) infection is thought to be a pathogenic contributor.

Outcomes: Clinical improvement was seen after first CHOEP with reduction in tumour mass. However her prognosis remained poor due to the presence of haemophagocytosis and resource-limited country of origin.

Conclusion: HIV co-infection should be considered in unexplained clinical presentations. This case highlights the ethical dilemma of provision of optimal health care to un-financial patients.

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