HIV and Cutaneous IgG4-rleated disease

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Background/purpose:
We report a case of cutaneous IgG4-related disease (IgG4-RD) in a 77 year-old male living with HIV, receiving suppressive antiretroviral therapy (ART) for over five years.

Approach:
In 2014 this man was diagnosed with HIV (CD4 count 16 cells/microL, HIV viral load 120,000 copies/ml) cryptococcal meningitis and an herpes simplex virus-2 ulcer in the left groin. He was treated with standard antifungal therapy, co-formulated abacavir/lamivudine/dolutegravir and acyclovir. Despite suppressive ART, he had a limited immunological response with a current CD4 count of 135 cells micro/L. The left groin ulcer relapsed in 2018 and was treated with valaciclovir. Despite treatment, he developed a large ulcerated, itchy exophytic mass 10x8cm in the left groin. A punch biopsy detected HSV-2 on PCR and histology showed lymphoplasmacytic infiltrate with predominantly plasma cells staining IgG4 positive with a conclusion of IgG4-related sclerosing disease. Additional stains showed no fungal/bacterial organisms and negative herpes simplex virus immunostaining. Further evaluation showed a markedly raised serum IgE level of 2,687 kunits/L (normal range 0-100), IgG4 6.20 g/L (normal range 0.04-0.86) and CRP 150 mg/L. Assessment for other manifestations of IgG4-RD did not identify any characteristic fibroinflammatory organ involvement either clinically or on CT scanning of the abdomen. MRI of the pelvis and upper legs revealed no regional lymphadenopathy or deep extension of the lesion

Outcomes/Impact:
Our patient was commenced on systemic corticosteroid therapy. A rapid decrease in his pruritus, lesion size and CRP was noted within a week.

Innovation and significance:
IgG4-RD is a reversible fibroinflammatory disease rarely described in association with HIV. Local HSV activation and IgG4-RDhave been reported in the context of eye lesions but not in extraocular locations. Our case highlights the importance of a tissue diagnosis in cutaneous lesions in individuals with persistently low CD4 counts on ART.

Disclosure of interest statement:
Nothing to disclose