Rare Tumours in HIV: A Case of EBV-Associated Smooth Muscle Tumour

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Background:
Epstein-Barr virus-associated smooth muscle tumours (EBV SMTs) are rare tumours of uncertain malignant potential. They predominantly affect immunocompromised hosts, including people living with human immunodeficiency virus (HIV).

Methods:
Case report (with written informed consent) and literature review.

Results:
A 27-year-old Indonesian man presented with two weeks of hiccups, headaches, fever and ataxia. He was diagnosed with advanced HIV (viral load 82,100 copies/mL, CD4 12 cells/µL) complicated by cerebral toxoplasmosis, cytomegalovirus viraemia and oral candidiasis. Antiretroviral therapy (bictegravir/emtricitabine/tenofovir alafenamide) and appropriate antimicrobials were initiated.

Computed tomography (CT) imaging performed for workup of opportunistic infections revealed multiple small “collections” in the right adrenal gland, spleen and left kidney. Biopsy of the adrenal lesion revealed a spindle cell proliferation positive by immunohistochemistry for smooth muscle actin and EBV-in situ hybridisation, consistent with EBV SMT. Bacterial, mycobacterial and fungal cultures were negative. EBV serology was positive (IgM-/IgG+) with low-level EBV viraemia (<1000 copies/mL).

Follow-up CT at 19 weeks of antiretroviral therapy with viral suppression and partial CD4 recovery (50 cells/µL) demonstrated enlarging adrenal and splenic lesions and new intra-abdominal lesions. Extensive investigation for opportunistic infection was negative. The progression was attributed to an immune reconstitution inflammatory syndrome (IRIS)-like response. The patient remains under radiological surveillance.

EBV SMTs are rare and our knowledge is limited to case series. They can affect any solid organ, and if multifocal, represent concurrent primary tumours rather than metastases. Surgical resection and immune reconstitution are the most described treatment approaches. While malignancy-associated IRIS has been described with Kaposi sarcoma and lymphoma, it is not defined in EBV SMT.

Conclusion:
EBV SMTs should be considered in the differential of mass lesions in patients living with HIV. This case highlights a hitherto undefined IRIS-like phenomenon in EBV SMT.

Disclosure of Interest Statement:
None.

AI Declaration:

AI has been used in the preparation of this submission, namely for language editing.