

Unusual case of HIV with Kaposi sarcoma and neuroendocrine tumour

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DESCRIPTION

A 50-year-old Black man with a 3-year history of HIV/AIDS on antiretroviral therapy (efavirenz/emtricitabine/tenofovir) presented with cough and dyspnoea. He had no significant medical history aside from HIV and had no history of opportunistic infections. He denied any history of smoking, alcohol or illicit drug abuse, and denied receiving blood transfusions. He had had unprotected sex with male partners in the past. His family history was positive for his mother having had a stroke. He had a history of non-compliance with antiretroviral therapy. Examination showed numerous brownish skin macules and purple coloured exophytic palatal lesions (figures 1 and 2). His CD4 count was 222 and viral load 1130 K copies/mL. Chest X-ray showed perihilar infiltrates (figure 3). CT of the chest revealed upper lobes and right middle lobe infiltrates mainly along the bronchovascular bundles



Figure 1 Brown macular skin lesion characteristic of Kaposi sarcoma.



Figure 2 Exophytic oral lesion characteristic of Kaposi sarcoma.



Figure 3 Chest X-ray showing perihilar infiltrates.

(figure 4). Bronchoscopy showed a polypoidal left lower lobe endobronchial lesion that was diagnosed as being Kaposi's sarcoma (KS). Transbronchial biopsies showed interstitial pneumonia and KS. The patient was categorised as having Centers for Disease Control and Prevention (CDC) stage C2 and WHO clinical stage 4 disease based on CD4 count and presence of KS. Over the next 2 years, the patient received several rounds of chemotherapy (paclitaxel, doxorubicin, imatinib and irinotecan) for KS, with improvement in pulmonary infiltrates. Unfortunately, he developed progressive abdominal pain and weight loss, and was noted to have an enlarging liver lesion that was a neuroendocrine tumour (NET), grade 2 (figure 5); he died shortly thereafter. Autopsy revealed widely metastatic NET.

KS is an angioproliferative disorder of endothelial origin that mainly affects mucocutaneous sites, but may also affect visceral organs. KS is

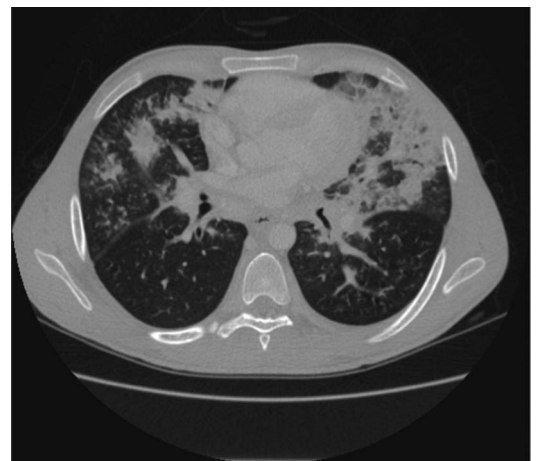


Figure 4 CT scan of the chest showing bilateral infiltrates.



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Figure 5 CT scan showing an enlarged liver completely filled with a tumour.

characterised by angiogenesis, the presence of spindle-shaped tumour cells, an inflammatory cell infiltrate dominated by mononuclear cells, extravasated erythrocytes, and oedema. KS is caused by human herpes virus 8 (HHV 8). HIV patients often develop pulmonary KS when CD4 counts are low.

Pulmonary KS can present as nodules, masses, infiltrates, effusions or purple/red endobronchial lesions. In the pre-antiretroviral therapy era, lymphoid interstitial pneumonia (LIP) and non-specific interstitial pneumonia (NSIP) were the most common HIV-related interstitial lung diseases.¹

Oral KS can involve the palate, tongue, gingiva, uvula and oropharynx as purple macules, patches, plaques, nodules or exophytic masses.² Cutaneous KS can present as purplish, reddish, brown or black macules, plaques or nodules.

NETs can arise from any organ system and are epithelial tumours with predominantly neuroendocrine differentiation. NETs, except for Merkel cell carcinoma and small cell lung cancer, are extremely rare in HIV. Less than 10 of NETs have been reported in HIV, the majority of them carcinoids.³ Our patient had a NET that was probably gastropancreatic in origin. To the best of our knowledge, this is the first reported case of an HIV patient who developed KS and a grade 2 NET.

Learning points

- ▶ Pulmonary Kaposi's sarcoma (KS) can present as nodules, masses, infiltrates, effusions or purple/red endobronchial lesions.
- ▶ Oral KS can involve the palate, tongue, gingiva, uvula and oropharynx as purple macules, patches, plaques, nodules or exophytic masses.
- ▶ Cutaneous KS can present as purplish, reddish, brown or black macules, plaques or nodules.

Competing interests None declared.

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