Case report

Skin nodules in a young patient with HIV/AIDS

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SUMMARY

AIDS-related Kaposi sarcoma (KS) is a malignancy seen in patients with HIV/AIDS that results from unrestrained human herpesvirus 8 infection. It can have an atypical presentation and an aggressive clinical course in patients with uncontrolled HIV infection. We present an interesting case of AIDS-related KS with an atypical initial presentation with skin nodules and debilitating lymphoedema. Patient was successfully managed with supportive measures, antiretroviral therapy and systemic chemotherapy.

BACKGROUND

Kaposi sarcoma (KS) is a low-grade vascular tumour associated with infection with human herpesvirus 8 (HHV-8), also known as the KS-associated herpesvirus. AIDS-related KS tends to have a variable clinical course, ranging from asymptomatic disease presenting as an incidental finding to a rapidly progressing neoplasm that can result in significant morbidity and mortality. The development of antiretroviral therapy (ART) has led not only to a remarkable decrease in the incidence of KS but also an improvement in the prognosis of patients with HIV-related KS.

CASE PRESENTATION

A 30-year-old man presented to the emergency department with painful swelling of his bilateral lower extremities and nodules on the skin. Symptoms started 2 months prior to the presentation. Physical examination revealed violaceous appearing firm and non-tender nodules on his chest, axilla and groin (figure 1), and 2–3+ pitting oedema on bilateral legs, thighs and scrotum. His past medical history was significant for HIV/AIDS diagnosed 5 years prior to presentation and he had history of non-compliance with ART. He is an active 1-pack/day smoker. His family history was unremarkable.

INVESTIGATIONS

Initial laboratory studies showed mild anaemia with a haemoglobin of 12 g/dL (normal range 13.5–17.5 g/dL) and a normal comprehensive metabolic panel. Further testing revealed a CD4 count of 35 cells/mm³ (normal range, 430–1276 cells/mm³) and a viral load of 55 000 copies/mL (normal range 0–100 copies/mL). Biopsy of one of the skin nodules revealed spindle-cell neoplasm arranged in whorls, which was positive for CD34 and HHV-8 on immunohistochemical testing.

DIFFERENTIAL DIAGNOSIS

Although violaceous appearing skin nodules in an immunocompromised patient raise strong suspicion for AIDS-related KS, possibility of other infectious and non-infectious aetiologies including bacillary angiomatosis, dermatofibromas, spitz nevus and hamartomas cannot be excluded based on clinical features only. Tissue biopsy with silver staining and immunohistochemical analysis is useful in ruling out other differential diagnoses.

TREATMENT

Once diagnosis was confirmed as AIDS-related KS, ART was resumed, and patient underwent systemic chemotherapy with liposomal doxorubicin 20 mg/m² every 3 weeks. 2D echocardiogram prior to initiation of chemotherapy revealed ejection fraction of 65%. He was followed in oncology and HIV clinics with clinical examination and laboratory studies every 3–6 weeks.

OUTCOME AND FOLLOW-UP

After receiving two cycles of liposomal doxorubicin along with ART (6 weeks of therapy), patient experienced complete resolution of the skin nodules and lower extremity oedema. He finished six cycles of chemotherapy and continues to be on ART with no recurrence of KS until.

DISCUSSION

With uncontrolled proliferation of HHV-8 in the setting of a compromised immune system, KS occurs 500–10 000-fold more commonly in HIV-positive individuals than in general population. Although occurring more commonly in patients with CD4 count less than 200 lymphocytes/mm³, CD4 count at presentation can be quite variable and KS may be the first manifestation of AIDS.¹

KS typically presents with cutaneous disease. Involvement of non-cutaneous sites like the oral cavity, gastrointestinal tract and respiratory system can be seen but visceral or osseous involvement as the initial presentation is relatively uncommon.² Clinical course tends to be highly variable from indolent lesions gradually shrinking over the years to a rapid increase in the number and size of lesions in some patients over the course of weeks to months, later being more commonly seen in patients who are non-compliant with ART.³ Despite the diagnosis often being clinical, biopsy should be performed to rule out other possibilities in an immunocompromised individual. Histological evaluation shows whorls of spindle-shaped cells with

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Figure 1 Photo of the chest showing violaceous appearing firm and non-tender nodules.

leucocytic infiltration and neovascularisation with aberrant proliferation of small vessels.⁴

If needed, immunohistochemical staining of biopsy specimens can be performed to detect the presence of HHV-8 latent antigen within the spindle cells, and PCR of the skin lesions can also be done to detect amplified HHV-8 DNA sequences, thus confirming the diagnosis.⁴

In addition to the well-defined lesions that are pathognomonic for KS, debilitating lymphoedema in the face, genitalia and lower extremities is often present. This comes as little surprise as KS is a neoplasm that originates in lymphatic endothelium, and thus, can disrupt the flow and permeability of the lymphatic system. Lymphoedema can have a significant impact on quality of life and requires early recognition and intervention with supportive measures and systemic chemotherapy to avoid serious debility.⁵ Patients with good-risk, asymptomatic KS with localised cutaneous involvement may be offered local treatment with intralesional chemotherapy and/or radiotherapy along with ART.⁶ Patient with poor-risk symptomatic visceral disease or rapidly progressive KS should be treated with ART combined with chemotherapy.⁷ Prognosis is largely determined by tumour burden and presence of systemic disease. Although visceral KS, especially gastrointestinal and pulmonary involvement may

prove fatal, chronic immunosuppression and opportunistic infections remain the major cause of morbidity and mortality in these patients.

Learning points

- ▶ Non-cutaneous presentation of Kaposi sarcoma with visceral or osseous involvement as the initial presentation is relatively uncommon and is typically seen in patients who are non-compliant with antiretroviral therapy (ART).
- ▶ Despite the diagnosis often being clinical, biopsy of accessible skin lesions should be performed to rule out other possibilities in an immunocompromised individual.
- ▶ Large volume symptomatic disease requires treatment with systemic chemotherapy in addition to ART.
- ▶ Adherence to ART with a low viral load and normal CD4 count is associated with good disease control and fewer recurrences.

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