Primary giant synovial sarcoma of the right ankle in a woman living with HIV-AIDS: a rare concurrence

Antonio Jose Reyes,¹ Wesley Greaves,² Kanterpersad Ramcharan,³
Fidel Rampersad⁴

¹Medicine, Infectious Disease Unit, San Fernando Teaching Hospital, San Fernando, Trinidad and Tobago

²Pathology, San Fernando Teaching Hospital, San Fernando, Trinidad and Tobago

³Medicine, Surgi-Med Clinic, San Fernando, Trinidad and Tobago

⁴Radiology, University of the West Indies, St Augustine, Trinidad and Tobago

Correspondence to

Dr Kanterpersad Ramcharan,
kramcharan79@yahoo.com

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DESCRIPTION

A 35-year-old woman living with HIV presented with progressive wasting syndrome and a slowly growing self-neglected right ankle mass for which she refused to seek medical attention and investigations (biopsy and image studies) for 12 months. There was no previous history of cancer, exposure to chemicals carcinogens or radiation, recreational drug use or alcohol consumption. Examination showed a body mass index of 18 kg/m² without oral thrush or lymphadenopathy. A large solid fungating dark mass of 26.0×20.0×18.0 cm in size, raised from the right ankle and extended beyond the skin openly. The severely painful tumour contained necrotic soft tissue alternating with haemorrhagic areas (figures 1 and 2). The mass effect on the right lower limb impaired ambulation. The rest of physical examination was normal.

The patient showed suppressed viral loads with good CD4+ Tcell counts (table 1). She also received highly active antiretroviral therapy (HAART) (table 2). Extensive investigations for other diseases were negative (table 1). MRI scan of the right ankle showed an irregularly shaped, infiltrating mass, measuring 1.0×3.8×3.1 cm³, located at the medial aspect of the right ankle, and extending into plantar region of the foot along the posterior tibial artery and nerve. This mass demonstrated low T1 signal, mildly high T2 signal and heterogeneous enhancement with intravenously administered Gadolinium. The tendons of foot appeared separate from the lesion. There was no osseous or articular involvement. The bones showed normal

Figure 2 Lateral view image showing the tumour extension beyond the right ankle including the heel and the skin and soft tissue surrounding the heel.

MRI signal (figures 3–5). CT scan of the chest revealed multiple soft tissue density bilateral lung nodules, that measured up to 1.0 cm in diameter, suggestive of pulmonary metastasis. Tumour biopsy with histopathology using routine H&E staining and immunohistochemistry supported the diagnosis of synovial sarcoma (figures 6–8). Diagnosed as primary right ankle synovial sarcoma stage IV with metastases to the

Figure 1 Posterior view image showing a large solid fungating dark mass of 26.0×20.0×18.0 cm in size, arising from the right ankle and extended beyond the skin openly. The mass contains necrotic tissue alternated with soft tissue bleeding areas.

Figure 3 MRI scan of right foot and ankle showed a 1.0×3.8×3.1 cm³ irregularly shaped lesion noted at the medial aspect of the right ankle and extending into plantar aspect of the foot, along the posterior tibial artery and nerve. The mass demonstrated high T2 signal on this sagittal fat saturation view.

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Images in...

Table 1 Medical investigations

Blood test	Result	Reference range
White cell count	8.7×10 ⁹ /L	4.5–11.0×10 ⁹ /L
Haemoglobin	77 g/L	117–155 g/L
Mean corpuscular volume	82.4 fL/red cell	80–96 fL/red cell
Platelets count	668×10 ⁹ /L	156–373×10 ⁹ /L
Venereal disease research laboratory Test	Non-reactive	Non-reactive or reactive
Fluorescent <i>Treponema pallidum</i> antibody absorption	Negative	Positive or negative
ELISA for HIV	Reactive	Non-reactive or reactive
HIV viral load on admission	Not detected	–
HIV viral load 2 months from admission	Not detected	–
CD4+ T cell count on admission	455 cells/μL	410–1590 cells/μL
CD4+ T cell count 2 months from admission	650 cells/μL	410–1590 cells/μL
Toxoplasma gondii IgG antibodies	0.43	Negative: less than 0.55
Toxoplasma gondii IgM antibodies	0.28	Negative: less than 0.55
Herpes virus 1 IgG antibodies	Negative	Index negative: Less than 0.9
Herpes virus 1 IgM antibodies	Negative	Index negative: Less than 0.9
Herpes virus 2 IgG antibodies	Negative	Index negative: Less than 0.9
Herpes virus 2 IgM antibodies	Negative	Index negative: Less than 0.9
Cytomegalovirus (CMV) IgG antibodies	0.700 UA/mL	Negative: Less than 1.5 UA/mL
CMV IgM antibodies	0.578 UA/mL	Negative: Less than 1.1 UA/mL
Epstein-Barr virus (EBV) IgG antibodies	Negative	Negative: Less than 0.8
EBV IgM antibodies	Negative	Negative: Less than 0.8
Hepatitis BS AG	Negative	Positive or negative
Hepatitis C IgG antibodies	Negative	Positive or negative
Hepatitis C IgM antibodies	Negative	Positive or negative
Pap smear	Normal	Normal or abnormal
Genital examination	Normal	Normal or abnormal
Mammogram both breast	Normal	Normal or abnormal
Both breast ultrasound	Normal	Normal or abnormal
CT scan of head, abdomen and pelvis	Normal	Normal or abnormal
CT scan of the chest	Abnormal	Normal or abnormal
MRI scan of the right ankle	Abnormal	Normal or abnormal

lung in an adult living with HIV-AIDS, she underwent right above knee amputation, and received palliative therapy. One month later, significant clinical improvement led to discharge her home and subsequent periodic attendant to the oncology and HIV outpatient clinics. Six months later, patient developed metastatic pulmonary haemorrhage and died from this non-aid defining cancer complication despite control of the HIV-AIDS.

In 2015, there were 36.7 million people living with HIV-AIDS worldwide but the global coverage of HAART

Table 2 Medical treatment

Intravenous therapy	Dosage	Period of treatment
Packed red blood cells	1 unit three times a week	2 weeks
Pethidine	100 mg twice daily	1 week
Dimenhydrinate	50 mg twice daily	1 week
Cefuroxime	750 mg three times daily	1 week
Subcutaneous drug	Dosage	Period of treatment
Enoxaparin sodium	40 mg	2 weeks
Oral drug	Dosage	Period of treatment
Efavirenz	600 mg daily	3 years
Tenofovir disoproxil/emtricitabine	245/200 mg per tablet 1 tablet daily	3 years
Pregabalin	150 mg daily	6 months
Cefuroxime axetil	500 mg twice daily	15 days
Tramadol hydrochloride/paracetamol	37.5/325 mg three times daily	15 days

reached only 46%. Several long-term studies have indicated an increased risk for the development of a multitude of non-AIDS-defining cancers in HIV-AIDS patients such as non-Hodgkin's lymphomas, Kaposi sarcoma and cervical cancer. Very few reports, however, exist regarding the occurrence of non-AIDS-defining sarcomas in the extremities.¹ The literature rarely mention the occurrence of bone and soft tissue tumours and also fail to describe the histological nature and site of these malignancies. Conventional synovial sarcoma among those patients with ages between 30 and 60 years is extremely rare. We have not found a previous publication regarding the occurrence of synovial sarcoma among

Figure 4 MRI scan of right foot and ankle shows low T1 signal on this T1-weighted sagittal view.

Figure 5 MRI scan of right foot and ankle with heterogeneous enhancement on IV gadolinium administration on this T1 weighted coronal view. It infiltrates into surrounding subcutaneous fat, without osseous or articular involvement. The tendons of foot appear separated from the lesion.

people living with HIV.¹ The pathogenesis of many malignancies associated with HIV infection is not yet completely understood, but it may involve concomitant viral or other

Figure 6 Tumour photomicrography: H&E staining with skin undermined by a monophasic synovial sarcoma with fascicles of spindle shaped cells without epithelial component (20× magnification).

Figure 7 H&E staining showing relatively ovoid uniform cells with occasional mitotic figures (200× magnification).

Figure 8 On immunohistochemistry, the tumour cells showed expression of Bcl-2 with strong staining and diffuse positive (4+) (40× magnification).

Learning points

- ▶ Synovial sarcoma of the ankle is an extremely rare and aggressive form of cancer but is perhaps unique as a concurrent malignancy among adults living with HIV-AIDS.
- ▶ Several comprehensive long-term follow-up studies rarely mention the occurrence of bone and soft tissue tumours among HIV-AIDS patients and also fail to describe the histological nature and site of these malignancies.
- ▶ The relationship of HIV-AIDS with osteosarcoma or sarcoma or synovial sarcoma remains uncertain
- ▶ The immunological basis of the development of malignancies in HIV-infected individuals is not yet completely understood. The pathogenesis of HIV inducing malignancy may involve direct virus carcinogenic actions or indirectly through immunosuppression.
- ▶ A greater index of suspicion is required in HIV patients presenting with unexplained bone and joint pain or swelling. Appropriate radiological investigation and timely referral to a specialised orthopaedic and oncology unit are recommended.

microbial infections with direct carcinogenic actions or indirectly through immunosuppression.^{2,3}

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