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## CASE REPORTS

# Kaposi sarcoma in an HIV-negative Tunisian patient: A rare cause of metatarsalgia

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### KEYWORDS

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**Abstract** *Background:* Kaposi sarcoma (KS) is an angioproliferative neoplasm that is commonly associated with human herpes virus-8 (HHV-8) and human immunodeficiency virus (HIV)/acquired immunodeficiency syndrome (AIDS). KS with osseous involvement is a rare occurrence, and is far more common in acquired immunodeficiency syndrome (AIDS)-related KS.

*Case presentation:* We present a 32-year-old Tunisian man, HIV negative, who presented with a 4-year history of atraumatic mechanical metatarsalgia that progressively worsened with a limping gait. Physical examination revealed marked symmetrical forefoot lymphedema and a painful restricted left knee joint movement. Physical examination showed purple-blue plaques and nodules on the feet and ankles. Serologic tests for HIV and syphilis were negative. Plain radiography of the feet revealed numerous small lytic lesions. There were also scattered lytic lesions in the metaphysis of the proximal tibia and fibula. Osteolysis was predominantly left. Magnetic resonance imaging of the feet showed abnormal bone marrow signal of metatarsals and phalanges. Skin lesion biopsy yielded the diagnosis of Kaposi sarcoma. The disease was managed with chemotherapy including vinblastine.

*Conclusion:* In a patient presenting with metatarsalgia without a commonly detected cause, it is mandatory to search for other lesions that may point to a rare diagnosis as KS which is famous for involvement of the metatarsal bone.

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## 1. Introduction

Kaposi sarcoma (KS) is a vascular neoplasm associated with Human Herpes virus-8 (HHV8) infection. Skin and mucous membranes are the most common sites, but other organs may be involved. Skeletal KS is rare and occurs either by direct spread of mucocutaneous lesions or through dissemination [1,2].

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There are 4 different clinical-epidemiologic variants of KS, including African (endemic) KS, classic KS, acquired immune deficiency syndrome (AIDS)-related (epidemic) KS, and transplantation (or immunosuppression)-associated KS. Prior to the AIDS epidemic, KS affected mostly cutaneous sites in African and European populations [3]. The American Cancer Society added a fifth type of KS; 'Kaposi sarcoma in human immunodeficiency virus (HIV) negative men who have sex with men'. KS with osseous involvement is rare, with less than 100 cases reported in the literature. In this article, we present a new case of non AIDS-related KS with bone involvement.

## 2. Case presentation

A 32-year-old male presented with a 4-year history of atraumatic mechanical metatarsalgia progressively worsened with a limping gait. Physical examination showed multiple, firm, purple-blue and well circumscribed plaques and nodules on the feet and ankles. He also had marked symmetrical lymphedema of the forefeet, and a painful restricted left knee joint movement. Laboratory analyses were normal. Plain radiography of the feet revealed regional osteopenia. Numerous small lytic areas were noted to result in a moth-eaten osseous lesion appearance. There were also scattered lytic lesions in the metaphysis of the proximal tibia and fibula. Osteolysis was predominantly left (Fig. 1).

Magnetic resonance imaging (MRI) of the feet was performed and showed abnormal bone marrow signals of metatarsals and phalanges (The signal intensity was decreased in T1-weighted images and increased in T2-weighted images/postgadolinium, suggesting bone marrow infiltration, edema and thickening of soft tissue (Fig. 2). Of these imagings, findings suggested the diagnosis of osteolytic bone metastases.

Therefore, a skin lesion biopsy was performed. Histological analysis showed a dermal nodular proliferation accompanied by perivascular infiltration of tumor spindle-shaped cells, frequent mitoses with Human Herpes virus-8 (HHV-8) positivity in immunohistochemistry. Diagnosis of KS was made. Serologic tests for HIV and syphilis were negative. Chest and abdomen computed tomography (CT) was normal. The disease was managed with chemotherapy including vinblastine.

## 3. Discussion

KS is an angioproliferative neoplasm. It is generally multifocal and most often involves the skin, with or without visceral involvement. Of the clinical-epidemiologic variants [4], our case is of the African endemic type. The sex ratio is variable and depends on the epidemiological form. African KS is more aggressive, typically presents with visceral involvement, lymphatic extension and is predominantly seen in young men of sub-Saharan Africa descent [5]. Despite the fact that HHV-8 infection results in the development of neoplastic disorders, such as KS in immunocompromised hosts, the pathogenesis of KS is still not clear [4]. Reported African patients with KS tested negative for HIV infection. Most patients with African KS originated from central Africa (predominantly Uganda in East Africa). Other patients were reported from Algeria and Morocco and from South Africa [3]. Cases of African-type Kaposi sarcoma involving the musculoskeletal system were mostly men mainly presenting in the metatarsals [6–9].

KS involvement of bone is unusual. Cortical bone erosion almost appears to represent local extension. It was previously recognized that osseous KS lesions are in general the result of contiguous invasion from a nonosseous lesion. Rarely, the starting point is intramedullary without adjacent skin lesion.



**Figure 1** Plain radiography of the feet showing numerous small lytic osseous lesions with a moth-eaten appearance.



**Figure 2** T1 weighted axial magnetic resonance imaging (MRI) of the feet showing abnormal bone marrow signal of metatarsals and phalanges. The signal intensity was decreased in T1 and increased in post-gadolinium, suggesting bone marrow infiltration, edema and thickening of soft tissue.

In this case, it is unclear whether these isolated osseous KS lesions represent primary musculoskeletal KS or a distant metastasis. In our case, the most likely mechanism of bone involvement is local extension. Classic and African KS affected most often the peripheral skeleton. Nevertheless, axial skeleton in AIDS-related KS is more likely involved and can lead to spinal cord compression [10]. Nodules and purplish, painless, sometimes ulcerated plaques of the limbs are characteristic findings in KS [11]. Up to 4.5% of osseous lesions may be detected on plain radiography in the classical and African endemic KS [3]. In a recent Moroccan study including 28 classic KS, bone involvement was the most frequent extracutaneous metastasis (28%). These authors reported pitting edema and functional impairment in nearly 25% of cases and a dramatic evolution in one patient with an above-knee amputation [12].

Although patients with osseous KS lesions are often asymptomatic, bone KS involvement can lead to bone pain, functional impairment and even pathologic fracture [3]. Bone involvement is almost always multifocal. However unifocal bone localization has been described: tibia, mandible, tarsus, and third phalanx [3,13]. Typically, topography of bone lesions is usually correlated with skin disease, and affected the extreme distality of the lower limbs (metatarsals, calcaneus, tibia, and fibula) [13]. Radiographic features of bone lesions KS are characterized by a well-defined area of osteolysis without peripheral bone condensation or sclerosis [13]. Bone pain may guide toward bone involvement in KS patients where further investigations using MRI may show local invasion of bone from cutaneous lesions and could be confirmed by bone biopsy [14]. CT-scan identified lytic bone lesions, but didn't distin-

guish them from other tumor, lymphoma or infectious lesions such as bacillary angiomatosis, main differential diagnosis. MRI appeared to be more sensitive for detecting the involvement of spongy bone and better demonstrates potential adjacent soft tissue lesions (hyposignal T1, increased signal in T2-weighted/post-Gadolinium MR images).

A pathological confirmation is mandatory for KS diagnosis. The biopsy findings often show vascular proliferation and spindle-shaped cells positive for the endothelial cell markers CD34 and HHV-8 positivity in immunohistochemistry.

It is now well defined that the mainstay of treatment of KS with bone involvement is based on systemic chemotherapy [4]. Local radiation therapy was recommended in some patients. Treatment of symptomatic lesions requires radiotherapy or chemotherapy. Similar to our case, a KS patient with bone pain was treated with vindesine, a synthetic derivative of vinblastine and the pain progressively disappeared [14].

*In conclusion*, in a patient presenting with metatarsalgia, without a commonly detected cause, it is mandatory to search for other lesions that may point to a rare diagnosis as KS which is famous for involvement of the metatarsal bone.

#### Conflicts of interest

No conflicts of interest.

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