Spinal Kaposi sarcoma presenting without cutaneous manifestations

Case report

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Kaposi sarcoma (KS) is one of the most common tumors in patients with human immunodeficiency virus (HIV), which characteristically presents with cutaneous lesions. The authors report a rare case of spinal KS with no cutaneous manifestation in a 32-year-old man with the acquired immunodeficiency syndrome who presented with abdominal pain. A computed tomography scan revealed incidental lesions in his lumbar spine, and additional imaging studies revealed numerous lesions in the lumbosacral spine and pelvis. An open biopsy was performed, and histopathological examination of the lesion confirmed the diagnosis of KS. At the time of presentation, the patient had no skin lesion or any other manifestation indicative of KS. The authors suggest that in HIV-positive patients who present with spinal lesions, KS should be included in the differential diagnosis. (DOI: 10.3171/SPI-07/11/558)

KEY WORDS • human herpesvirus 8 • human immunodeficiency virus • Kaposi sarcoma • spine

Abbreviations used in this paper: AIDS = acquired immunodeficiency syndrome; CT = computed tomography; HHV8 = human herpesvirus 8; HIV = human immunodeficiency virus; KS = Kaposi sarcoma; MR = magnetic resonance.
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tion. Motor and sensory examinations yielded normal find-
ings. He reported no radicular pain, numbness, tingling, or
weakness.

To evaluate his abdominal symptoms, a CT scan of the
abdomen and pelvis was obtained. The internal organs
were normal, but incidental hypodense lesions were seen in
the sacrum and several vertebral bodies of the lumbar spine
(Fig. 1). Magnetic resonance imaging studies of the lumbo-
sacral spine showed hypointense lesions on T1-weighted
imaging that enhanced intensely after the administration of
gadolinium and were hyperintense on T2-weighted MR
imaging (Fig. 2). The differential diagnosis at this time was
infectious instead of congenital hemangiomas. On comple-
tion of a course of intravenous antibiotics, follow-up MR
imaging studies showed progressive growth of these lesions.
The differential diagnosis at this stage was a neoplastic (mul-
tiple myeloma, lymphoma) or infectious (tuberculosis, sar-
coidosis, bacterial, fungal) process. A bone marrow biopsy
was then performed and showed no evidence of disease.

Operation. A CT-guided needle biopsy initially perform-
ed on one of the sacral lesions yielded nondiagnostic re-
sults. The Neurosurgery Service was consulted for an open
biopsy procedure that was performed on the right L-4 pedi-
cle lesion.

Pathological Findings. The biopsy specimen showed frag-
ments of bone and fibrous tissue that contained a tumor
composed of a monomorphic proliferation of spindle cells
forming small vessels with irregular contours. Endothelial
proliferation was present (Fig. 3A), and the tissue stained
positive for HHV8 (Fig. 3B).

Postoperative Course. The patient was hospitalized over-
night and was discharged the following day on oral anal-
gesics. In addition to the highly active antiretroviral therapy
that he was taking previously, he was started on vincristine
biweekly as recommended by the Oncology Service. Three
months later, follow-up MR imaging showed minimal re-
gression of the lesions. His viral load was still stable, and
clinical examination showed that he had gained weight.

Discussion

Kaposi sarcoma is a mesenchymal tumor that was first
described by Dr. Moritz Kaposi in 1872. It was considered
a rare tumor of the skin until the HIV epidemic occurred,
and the incidence of the disease increased strikingly in the
immunocompromised population, with 15 to 20% of HIV-

Fig. 1. Axial CT scan of the lumbar spine obtained without
addition of contrast material, revealing an intravertebral hypodense
lesion (arrow).

Fig. 2. Sagittal T1-weighted MR images of the lumbar spine without (A) and with (B) addition of gadolinium demonstrat-
ing an intravertebral enhancing lesion (arrows) at the L-4 level. Axial T2-weighted MR image (C) demonstrating an
intravertebral hyperintense lesion (arrow) at L-4.
infected men developing KS. Internal organs, particularly the gastrointestinal tract, lungs, and lymph nodes, have been reported as primary sites of KS. Spine involvement has been documented, and in fact, spinal decompression was performed to alleviate symptoms of spinal cord compression. However, these lesions were a later manifestation after the initial appearance of the disease in different organs (Table 1).

The hallmark of KS is the characteristic skin lesion that is seen in most cases. It is unusual to see the clinical presentation of KS in the spine. In fact, when the disease first became more prevalent in HIV-infected patients, the literature seemed to indicate that KS was not a disease of the spine. Vertebral lesions in a patient with cutaneous lesions seemed to support the differential diagnosis of bacillary angiomatosis over KS. We now describe what we believe is the first reported case of KS in the spine without manifestation of the disease in the skin or in any other location such as the lungs or the gastrointestinal tract.

In recent years, it has been shown that HHV8 is necessary but not sufficient for the development of KS. In all likelihood, there are probably genetic and environmental factors in addition to HHV8 that cause KS.

**Treatment Modalities**

Therapeutic modalities for KS depend on the extent of the disease. Highly active antiretroviral therapy is an effective prophylactic and therapeutic regimen for AIDS-related KS. In cases of transplant-related KS, the recommended approach is modification of immunosuppressive regimens to the lowest dose consistent with graft function, with a response rate close to 50%. Systemic chemotherapy is generally reserved for patients with visceral or progressive KS. A variety of therapeutic modalities has been used, including the administration of liposomal doxorubicin, liposomal danorubicin, vincristine, etoposide, cisplatin, interferon, vinblastine, bleomycin, paclitaxel, and bleomycin/vincristine in combination, with success rates between 59 and 88%. Localized and cutaneous lesions can be managed with local therapies such as radiotherapy, cryotherapy, cryosurgery, and laser surgery; however, radiotherapy has been related to an increased long-term risk of developing carcinomas.

**Natural Course**

Regression has been observed in transplant-related KS after the dose of immunosuppressive treatment is lowered;

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**TABLE 1**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Patient Age (yrs), Sex, HIV Status</th>
<th>Symptoms</th>
<th>Primary KS</th>
<th>Lesion Location</th>
<th>Management</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Isenbarger &amp; Aronson, 1994</td>
<td>29, M, HIV+</td>
<td>hemoptysis</td>
<td>skin, lungs</td>
<td>multiple undiagnosed thoracic spine lesions on chest CT scan</td>
<td>autopsy, vertebral lesions were identified as KS</td>
<td>died of pulmonary hemorrhage</td>
</tr>
<tr>
<td>Turner et al., 1997</td>
<td>33, M, HIV+</td>
<td>back pain</td>
<td>skin</td>
<td>multiple lumbar lesions</td>
<td>open biopsy, bleomycin biweekly for 6 mos</td>
<td>skin lesions receded; bone lesions unchanged on imaging studies</td>
</tr>
<tr>
<td>Prieto, 1998</td>
<td>34, M, undetermined</td>
<td>back pain</td>
<td>lung</td>
<td>L-3 vertebra</td>
<td>open biopsy, supportive care</td>
<td>died in hospital of pulmonary hemorrhage</td>
</tr>
<tr>
<td>van Twillert et al., 2004</td>
<td>33, M, HIV+</td>
<td>paraplegia</td>
<td>skin, GI</td>
<td>T-10 &amp; L-2 vertebrae &amp; epidural space</td>
<td>open biopsy &amp; radiotherapy</td>
<td>refused further therapy; died of pneumonia</td>
</tr>
<tr>
<td>55, M, HIV+</td>
<td>55, M, HIV+</td>
<td>paraplegia</td>
<td>skin, GI</td>
<td>T-5 vertebra &amp; epidural space</td>
<td>laminectomy &amp; decompression</td>
<td>died of pulmonary hemorrhage</td>
</tr>
</tbody>
</table>

* GI = gastrointestinal.
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it occurs spontaneously in 2 to 20% of classic KS cases and rarely in AIDS-related KS.11 The latter disease regresses after effective highly active antiretroviral therapy or chemotherapy. The AIDS-related form of KS has been staged according to the AIDS Clinical Trials Group classification system, with recent modification since the use of highly antiretroviral therapy began.10 Patients are usually classified as having good or poor risk based on their tumor burden, immune function as measured by CD4 T-lymphocyte count, and the presence of systemic illness. Multivariate analysis demonstrated that the combination of poor tumor stage and systemic disease identified patients with a poor prognosis, with a 3-year survival rate of 53%, compared with 80% in patients without this combination.

Conclusions

Kaposi sarcoma may not present with typical findings in HIV-positive patients. Our case of KS is exceptional because of its presentation as spine lesions alone. This case shows that lesions may occur without a cutaneous manifestation in an area not typically associated with the disease, and thus a diagnosis of KS cannot be ruled out by a rare location, specifically that of the spine. Thus, neurosurgeons need be aware of its presentation. Furthermore, it is important to take into account the risk factors associated with KS, which include but are not limited to the patient’s HIV status, sex, sexual preference, and ethnicity. We suggest that the differential diagnosis of spine lesions in patients with HIV, even if there is no cutaneous manifestation, should include KS.

References


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