EXTRA-THORACIC KAPOSI'S SARCOMA: EVIDENCES OF A MULTISYSTEMIC INVOLVEMENT

Dr. Javier Willatt H(1), Int. Cristian Moyano N(2), Int. Camilo Apey R(2), Dr. Leonardo Lidid A(3).

1. Radiology Resident. West Campus, School of Medicine, Universidad de Chile, Hospital San Juan de Dios.
2. Medical Intern. West Campus, School of Medicine, Universidad de Chile, Hospital San Juan de Dios.
3. Assistant Professor of Radiology. West Campus, School of Medicine, Universidad de Chile, Hospital San Juan de Dios.

Abstract

Kaposi's sarcoma is a low-grade vascular neoplasm that typically manifests as one of four variants, being the epidemic form (or AIDS-related variant) the best known of them four. The pulmonary involvement of this disease has been well described and it is expressed mainly by bilateral parenchymal nodules of characteristic morphology; however, this tumor produces a multisystemic compromise not sufficiently known. Due to the increasing incidence and survival rates of patients with HIV, and the fact that Kaposi's sarcoma is one of its frequent complications, we sought to describe its staging and major imaging findings on computed tomography scans, given the importance of timely recognition of this neoplasm for a proper prognosis and management of the disease.

Keywords: Neoplasm staging, liver, Kaposi's sarcoma, AIDS, musculoskeletal system, gastrointestinal tract.

Introduction
Kaposi’s sarcoma is a low-grade mesenchymal tumor that involves blood and lymph vessels, primarily affecting the skin, which can cause disseminated disease in various organs \(^{(1)}\).

There are four types of the disease, differentiated by the kind of patients each one of them affect and the severity of patients clinical picture \(^{(2)}\): classical, endemic (African), epidemic (associated with AIDS), and iatrogenic (transplant-related) variants.

In AIDS patients, this pathology is a common complication and currently the main cause of death, according to some international series, thus displacing opportunistic infections. This change seems to be conditioned by the rapid and efficient recovery of the immune system achieved by the introduction and widespread use of active antiretroviral therapy, which has expanded globally in recent years \(^{(3)}\).

In Chile, there are no clear official data of the prevalence of Kaposi’s sarcoma or its treatment. The incidence rate of this disease at Hospital Dr. Sótero del Río (Santiago, Chile) rose from 0.47 cases per 100 person-years (1996) to 1.52 cases per 100 person-years (1999) \(^{(4)}\). In a study carried out in 1998 at Hospital Salvador (Santiago, Chile), 6.27% of AIDS patients seen at that institution had Kaposi's sarcoma \(^{(5)}\).

Due to the above, it is important that the radiologist knows the spectrum of presentation of this disease, that may involve multiple organs and systems, in which the computed tomography (CT) diagnostic imaging plays a fundamental role by providing the necessary information for the proper prognosis and management of these patients.

Therefore, we decided to review the staging of Kaposi’s sarcoma and its imaginological manifestations, both the most significant and the least known, as exemplified by imaging studies of cases that have occurred at San Juan de Dios Hospital.
Staging

Like all neoplasias, this disease has its own staging and, although there is no consensus on a shared framework for defining prognosis, most commonly used criteria are the ACTG (AIDS-Clinical Trial Group). Due to the widespread use of active antiretroviral therapy, there are recent proposals for modification of these criteria (3) consisting of the elimination of CD4 level as classification category, leaving only the tumor extension (T) and the systemic state of the patient (S) as criteria to be used (Table I).

In this area, the main task of the radiologist is to establish the extent of tumor (T), determining presence or absence of visceral involvement, as well as identify and evaluate the existence of opportunistic infections (S). Although skin lesions occur in relatively early stages of the evolution of patients with AIDS, the commitment of multiple sites and organs may occur as a late phenomenon, with CD4 counts less than 200 cells mm$^3$ (6). In this context, it should be noted that pulmonary neoplastic involvement corresponds to one of the visceral commitments with worse prognosis in the evolution of this disease (3).

In autopsies of patients with AIDS, visceral commitment in order of frequency is: lymph nodes (72%), lung (51%), gastrointestinal tract (48%), liver (34%), and spleen (27%); musculoskeletal system involvement is much rarer (7,8). Interestingly, the visceral involvement may not be accompanied by skin lesions in up to 29% of cases (9).

Transplant patients have Kaposi’s sarcoma in up to 5.7% of cases, with a media of 21 months for development of disease, according to reports of some publications (1). In these patients, a different staging is applied, although the role of the radiologist is quite similar (Table II) (10).

Areas of engagement
**Lymphadenopathy**

The presence of lymphadenopathy in AIDS patients may be secondary to various causes, among which we can cite: Kaposi’s sarcoma, HIV-associated lymphoma, generalized hyperplasia or mycobacterium infection (tuberculosis or avium) \(^{(11)}\). On phase contrast CT, the demonstration of hyperdense adenopathies is highly suggestive (but not exclusive) of Kaposi’s sarcoma (Figure 1), although the absence of this finding does not rule out the diagnosis since it also can present with iso- or hypodense lymphadenopathy \(^{(12)}\).

---

**Table I. Staging of Kaposi’s sarcoma related to AIDS (modified ACTG) \(^{(3)}\)**

<table>
<thead>
<tr>
<th>Tumor extension (T)</th>
<th>Patient’s clinical state (S)</th>
</tr>
</thead>
<tbody>
<tr>
<td>T0: Good prognosis</td>
<td>S0: Good prognosis</td>
</tr>
<tr>
<td>T1: Poor prognosis</td>
<td>S1: Poor prognosis</td>
</tr>
</tbody>
</table>

- **T0:** Good prognosis: Confined to skin and/or lymph nodes and/or minimum commitment of oral cavity.
- **T1:** Poor prognosis: Extensive oral commitment, presence of edema, ulcer, tumor, or gastrointestinal disease, or other.
- **S0:** Good prognosis: No history of opportunistic infection or B symptoms (fever or unexplained night sweats, unexplained weight loss of 10% or persistent diarrhea) and Karnofsky score $\geq 70\%$.
- **S1:** Poor prognosis: History of opportunistic infections, symptoms B, other HIV-related diseases and Karnofsky score $<70\%$.

---

**Table II. Staging of Kaposi’s sarcoma in transplant patients \(^{(10)}\).**

<table>
<thead>
<tr>
<th>Stage 1</th>
<th>Localized skin lesions involving just one limb</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage 2</td>
<td>Extensive skin lesions involving more than one limb.</td>
</tr>
<tr>
<td>Stage 3</td>
<td>Generalized: visceral involvement and/or lymph nodes and/or skin</td>
</tr>
<tr>
<td>--------------</td>
<td>---------------------------------------------------------------</td>
</tr>
<tr>
<td>Stage 4</td>
<td>Any of the above in the presence of infections or other life-threatening associated malignancies</td>
</tr>
</tbody>
</table>

**Chest**

The commitment of the chest occurs in approximately 45% of cases, while in 15% of cases absence of cutaneous involvement can be observed\(^{(13)}\). It may affect: pulmonary parenchyma, trachea, lymph nodes, pleura, and chest wall, with or without opportunistic infection.

On plain radiography, this condition can manifest as a reticular pattern associated with pulmonary nodules, although pathological findings occasionally may not be found with this method of study. On CT explorations, the presence of poorly defined bilateral nodules, > 1 cm in diameter, symmetrical, peribronchovascular distribution, and with "in flame" morphology, are characteristic and highly suggestive (Figure 2)\(^{(14)}\). However, there is another broad spectrum of nonspecific pulmonary manifestations ranging from septal thickening, nodules, areas of ground glass, condensation, and cavitation, to the existence of pleural effusion and collections, making its appearance often indistinguishable from lymphoma, *brachogenic carcinoma* or lung infection, including among others *Pneumocystis jiroveci pneumonia*, and bacillary angiomatosis (*Bar-Tonella henselae* infection)\(^{(1)}\).

**Liver and spleen**

The ultrasound study may show hepatomegaly, with heterogeneous parenchyma and small multifocal hyperechoic nodules (5-12 mm in diameter) usually located in periportal position. CT explorations may not demonstrate alterations or only the existence of hepatosplenomegaly\(^{(11)}\); however, small
hypodense nodules associated with hilar and peripheral portal branches growth are also described. These nodules are most evident in the phase contrast and many of them have delayed enhancement (4-7 min), appearing as iso- or hyperdense, being indistinguishable from hemangiomas or peliosis; similar findings are described in the spleen (1, 15). Other differential diagnoses to consider for small hypodense lesions in these organs are bacillary angiomatosis and microabscesses (11).

Figure 1. A 40-year-old patient with AIDS and gastric Kaposi’s sarcoma. a) Adenopathy of the greater curvature (arrow) that captures contrast is observed. b) Greater impregnation of the adjacent anterior rectus muscle must be noted
Figure 2. Pulmonary involvement by Kaposi’s sarcoma. a) Scout view in a patient with severe compromise, showing multiple nodules in both lungs. b) CT scan of another patient, carrier of penile Kaposi’s sarcoma with poorly defined multiple bilateral pulmonary nodules, greater than 1 cm in diameter, symmetrical, with peribronchovascular distribution, “in flame ” morphology.

Gastrointestinal tract

Gastrointestinal tract involvement is one of the most common visceral extensions in disseminated disease, reaching 50% of these patients in some series (1). Involvement may extend from the oropharynx to the rectum, including the gallbladder; the duodenum is the portion of the digestive tract that most often gets involved (16).

Baritated studies play no role in the detection of early flat lesions, although in larger lesions may show filling defects with or without central ulceration (bull’s eye).

On CT scans, polypoid submucosal masses are 0.5 to 3 cm in diameter and irregular thickening of the folds may be found. They may appear associated with regional lymphadenopathy presence as well as with other lymph node groups (Figure 3). Due to this characteristic submucosal location, endoscopic biopsies may be negative, a factor to be considered when evaluating the possibility of other studies in these patients (1).
Figure 3. Gastric wall thickening in patient of Figure 1, whose biopsy corresponded to Kaposi’s sarcoma. The patient had no obvious skin lesions at the time of performing CT scan.

**Genitourinary tract**

The skin of the penis is mainly affected (1, 17) (Figure 4). The commitment of the adrenal glands, bladder, scrotum, seminal vesicles and testes is very rare. Hydronephrosis and obstructive uropathy secondary to retroperitoneal lymph nodes growth may be found (18).
Figure 4. *Kaposi’s sarcoma of the penis, same patient as in figure 2b. The visual appearance of the lesions on physical examination is characteristic, although the bacillary angiomatosis can emulate their morphology."

**Musculoskeletal System**

Musculoskeletal system involvement is rare and usually (but not only) is secondary to extension due to proximity with adjacent skin lesions. Muscular commitment is generally due to contiguity from subcutaneous cellular tissues, causing edema and a soft tissue mass without specific features, findings especially demonstrable with magnetic resonance imaging (MRI), where it is possible to observe a marked contrast uptake because of the vascular nature of the injury (1, 19). In the osseous system, the axial and appendicular skeleton are usually affected by edema and cortical lesions ranging from erosion to overt destruction, rarely associated with periosteal reaction; these findings may be observed on both plain radiography and CT projections, while osseous edema is shown on MRI sequences (20, 21).

The main differential diagnoses are: lymphoma, tuberculosis and, particularly, bacillary angiomatosis; biopsy is required to obtain the definitive diagnosis. However, it is described that a positive thallium scintigraphy in conjunction with a negative gallium scintigraphy (usually positive in lymphoma and bacillary...
angiomatosis) is considered highly suggestive of Kaposi’s sarcoma \(^{(19, 21)}\).

**Conclusions**

Kaposi’s sarcoma is a well-known complication of AIDS and transplant immunosuppression, which can even cause death. The main role of the radiologist is to establish its diagnosis and determine the existence of visceral involvement, highlighting pulmonary nodules of "*in flame*" morphology as the most characteristic features for diagnosis; in the rest of the economy a number of nonspecific findings are observed, commonly difficult to differentiate from other entities without an histological study.

Bacillary angiomatosis is the differential diagnosis that best mimics the Kaposi’s sarcoma pattern of extrathoracic commitment, even in the visual appearance of skin lesions (Figure 5). Other differential diagnoses worthy of being kept in mind are the following: opportunistic infections, tuberculosis, and lymphoma. Accordingly, it is always necessary to consider patients history and application of other methods of study for a more precise diagnosis.
**Figure 5.** Same patient as in Figure 2b with pulmonary Kaposi’s sarcoma. Some months after the diagnosis he experienced a significant proliferation of characteristic pulmonary nodules associated with liver damage a), spleen lesions b) and bone damage (c, d). This was interpreted as progression of his baseline sarcoma. Extrapulmonary lesions mentioned were identical to that of bacillary angiomatosis, diagnosis that could not be discarded with absolute certainty.
References


