More Than Skin Deep

What abdominal radiologists need to know about AIDS associated Kaposi sarcoma

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Disclosures

• *Neither authors nor their immediate family members do not have a financial relationship with a commercial organization that may have a direct or indirect interest in the content.*
Outline

• Review of pathophysiology
• Background of Kaposi sarcoma (KS) in AIDS
• Overview of KS in abdominal organs
  – Imaging findings can be non-specific, need to have a high-level of suspicion in patients with AIDS
  – Review of GI, liver, lymph node, soft tissue and osseous manifestations
• Take home points
What is Kaposi sarcoma (KS)?

- Multicentric angioproliferative neoplasm that can involve skin and solid organs\(^1,2\)

- Arises from native endothelial cells found throughout the body\(^3\)

- Can also metastasize

H+E: Spindle cells, vascular channels and extravasated red blood cells

Nuclear and granular staining of HSV8 (seen in 100% of KS)
Human Herpes Virus 8 infects endothelial type KS progenitor cells, leading to malignant transformation and the formation of KS spindle cells.
Types of KS

- **Mediterranean** – uncommon, primarily occurs in middle aged to elderly men

- **Endemic** – common in young men from equatorial Africa

- **Transplant related** – thought to be secondary to immune suppression

- **Epidemic or HIV-related** – the most common neoplasm in AIDS and the most prevalent form (> 90%) of KS in N. America. This subtype is the focus of our exhibit.\(^1,^4\)
The widespread adaptation of cART (combined antiretroviral therapy) has lead to dramatic reduction in AIDS associated Kaposi’s sarcoma (KS)
What is the impact of KS in AIDS?

- Nearly 500,000 Americans are currently living with AIDS\textsuperscript{5}
- Many are not under active therapy and thus at high risk for KS
- KS can involve any organ. Up to 30% of patients with KS will not have any skin involvement
- In an autopsy series, the most common sites of visceral KS were lymph nodes (72%), GI tract (48%), liver (34%), and spleen (27%)\textsuperscript{6}
Gastrointestinal Tract

- Can involve anywhere along the GI tract, affects up to 40% of KS patients\(^6\)

- Manifests as irregular thickening or polypoid mass, which can ulcerate

- Usually asymptomatic; however can present with abdominal pain, weight loss, nausea, emesis, malabsorption, or diarrhea

- Severe complications include intussusception, bowel obstruction, bleeding, appendicitis, and perforation\(^7,8\)
KS nodules scattered through the GI tract
Colo-colonic intussusception due to KS nodule
Circumferential GI wall thickening is a common manifestation of KS. Circumferential rectal wall thickening, perirectal stranding and adenopathy. Location can be easily sampled via endoscopy or endoscopic ultrasound. Surgical biopsy was definitive for KS.
Fluoroscopy findings in the GI tract

- Submucosal masses and irregular folds as seen in the stomach on the right

- Ulcers can also demonstrate a classic target or bull’s eye appearance

Adapted from Boland G et. Al Gastrointestinal Imaging: The Requisites 4th edition 2013, p 65
Hepatic KS

- Liver involvement can be seen in up to 35% of AIDS patients\(^6\,\text{,}^{10}\)

- Clinical presentation: abdominal pain, elevated transaminases, hypoalbuminemia, elevated alkaline phosphatase, and hepatomegaly

- Imaging: Hepatomegaly, or infiltrating soft tissue / nodules along a perivascular distribution\(^{10}\)
CT demonstrates hypoattenuating periportal soft tissue. Differential includes periportal edema; however this patient's disseminated KS and lesion chronicity over two years favored hepatic KS.

Gross pathology example: Reddish irregular masses in a periportal infiltration.
Multifocal Hepatic KS nodules

Coronal and Axial T2 images show T2 hyperintense nodular and infiltrative perivascular involvement of KS

The patient was 33 y/o HIV+ M who p/w severe abdominal pain, hemoptysis (with radiographic findings of pulmonary KS), and fever
KS can manifest as perivascular soft tissue mass

• Since KS arises from native endothelial precursors, it also commonly presents as a perivascular mass

• In one series, up to 50% of patients had perivascular thickening on pulmonary imaging\textsuperscript{11}

• Abdominal perivascular involvement is not well described in the literature, however we have seen several cases
Perivascular involvement along the mesenteric vasculature
Perivascular soft tissue infiltration along the femoral vessels
Lymph Nodes

• LN enlargement is common and nonspecific in AIDS

• Differential includes: HIV related lymphadenopathy, TB, syphilis, lymphoma, infections, and other malignancies

• Clinical presentation: usually asymptomatic, but symptoms also include classic B-cell symptoms like night sweats, fevers, and weight loss\(^{12}\)
Bulky lymphadenopathy can be evident on visual inspection and is associated with an aggressive course. The enlarged lymph nodes above lack a normal fatty hilum and some demonstrate central necrosis.
Hepatic and LN involvement

- Geographic subcapsular periportal soft tissue and confluent areas in the right hepatic lobe measuring up to 3 cm

- Multiple enlarged bilateral inguinal lymph nodes, which lack a normal fatty hilum and some contain areas of central necrosis
Musculoskeletal

• Uncommon manifestation

• Can infiltrate bone, marrow, and skeletal muscles
  – Most from direct infiltration from adjacent organ
  – Also lytic lesions with non-sclerotic borders

• Can present clinically with limited mobility, bony or muscular pain$^{13,14}$
Musculoskeletal

Bony lytic lesions measuring up to 9 mm. This patient had extensive disseminated KS with lesions found through the lymph nodes, axial skeleton, and perirectal tissues. This is a typical appearance for KS, which can also erode through the bony cortex.
• KS involvement of the labia: nodular and swath-like thickening with avid enhancement, as well as subcutaneous soft tissue nodule
Other organs

- **Spleen**
  - Rare
  - Imaging appearance is similar to liver: nodular and infiltrative soft tissue

- **Renal**
  - Very rare
  - Can involve anywhere from the kidneys, down the ureters, to the bladder
  - Clinical presentation: hydronephrosis (2/2 to obstructing masses), hematuria\textsuperscript{15}
Clinical Course of Systemic KS

• Systemic KS is often progressive and severe

• Common source of vague abdominal pain or obstruction

• Significant cause of morbidity and mortality

• Often concomitant pulmonary involvement, which can manifest as dyspnea, SOB, or respiratory failure$^{4,11}$
Treatment

- Only 70% of patients respond to cART alone
- Systemic KS often necessitates the addition of chemotherapy
- No significant difference in outcomes between common chemotherapy regimens which include doxorubicin, daunorubicin, and paclitaxel.\textsuperscript{16}

Figure obtained from the NIH
http://ehp.niehs.nih.gov/117-a75/
• AIDS-related KS is the most common form in America accounting for 90% of cases

• Up to 30% of KS patients can lack any skin involvement, thus the radiologist can play a crucial role in identifying clinically occult disease
  – GI tract: nodular thickening anywhere along GI tract
  – Liver: hepatomegaly; periportal soft tissue
  – Perivascular soft tissue along mesenteric or iliac vessels
  – Bulky lymphadenopathy, can be avidly enhancing +/- necrosis
  – Lytic bone lesions
  – Dermal and subcutaneous thickening and nodularity

• Knowledge of typical KS findings and inclusion in the differential in patients with AIDS can aid in timely diagnosis and appropriate management
References


Thank you

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