



Hidden disseminated extracutaneous AIDS-related Kaposi sarcoma

Naoki Kawakami^a, Ho Namkoong^{b,c,*}, Masayuki Shimoda^d, Hiroshi Kotani^{e,f},
Hiroshi Fujiwara^e, Naoki Hasegawa^e

^a Department of Emergency and Critical Care Medicine, St. Luke's International Hospital, 9-1 Akashicho, Chuo-ku, Tokyo 104-8560, Japan

^b Division of Pulmonary Medicine, Department of Medicine, Keio University School of Medicine, 35 Shinanomachi, Shinjuku-ku, Tokyo 160-8582, Japan

^c Laboratory of Clinical Immunology and Microbiology, National Institute of Allergy and Infectious Diseases, NIH, Bethesda, MD, 20852, USA

^d Department of Pathology, Keio University School of Medicine, 35 Shinanomachi, Shinjuku-ku, Tokyo 160-8582, Japan

^e Center for Infectious Diseases and Infection Control, Keio University School of Medicine, 35 Shinanomachi, Shinjuku-ku, Tokyo 160-8582, Japan

^f Department of Pharmacy, Keio University School of Medicine, 35 Shinanomachi, Shinjuku-ku, Tokyo 160-8582, Japan

ARTICLE INFO

Article history:

Received 25 January 2020

Received in revised form 6 February 2020

Accepted 6 February 2020

Keywords:

Disseminated Kaposi Sarcoma (KS)
Human immunodeficiency virus (HIV)
Acquired immune deficiency syndrome
(AIDS)

ABSTRACT

A 68-year-old man with past medical history of multiple cerebral infarctions presented to our hospital with subacute paresis. His vital signs on presentation were normal, and his physical examination, other than his neurological findings, was unremarkable. Neurological examinations suggested cerebellar ataxia. Laboratory testing confirmed positive for human immunodeficiency virus (HIV) infection. His CD4-positive lymphocyte count was 45/ μ L, and HIV-RNA was 2.3×10^5 copies/mL. Brain computed tomography (CT) scan revealed multiple mass lesions and brain magnetic resonance imaging (MRI) with fluid-attenuated inversion-recovery (FLAIR) revealed periventricular hyperintensities, which suggested multiple malignant lymphoma and HIV encephalopathy. His state of consciousness had gradually worsened. Eventually, he died one month after admission. The autopsy unexpectedly showed disseminated Kaposi's sarcoma (KS). KS lesions were found in the stomach, small intestine, liver, spleen, mesentery and lungs. KS was not observed on his skin. Gross findings revealed multiple nodular lesions in each organ, and hematoxylin and eosin staining showed proliferation of spindle cells with vascular proliferation. Immunostaining was positive both for endothelial marker (CD31 and von Willebrand factor) and lymphatic endothelial marker (D2-40), which were consistent with KS. KS is the most common tumor in AIDS patients. It is caused by the human herpes-virus 8 infection. It manifests an indolent clinical course and mostly involves cutaneous lesions over the lower limbs, trunk and oral cavity. In this case, autopsy revealed disseminated KS pathologically, which was unrecognized before his death. This case highlights the possible existence of disseminated KS even without its cutaneous findings.

© 2020 Published by Elsevier Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

Case illustrated

A 68-year-old man, a former smoker, with past medical history of multiple cerebral infarctions presented to our hospital with subacute paresis. He was a man who had sex with men (MSM). His vital signs on presentation were normal, and his physical examination, other than his neurological findings, was unremarkable. Neurological examination suggested cerebellar ataxia. Laboratory testing was positive for human immunodeficiency virus (HIV) infection. His CD4-positive lymphocyte count was 45/ μ L, and HIV-RNA was 2.3×10^5 copies/mL. Brain computed

tomography (CT) scan revealed multiple mass lesions and brain magnetic resonance imaging (MRI) with fluid-attenuated inversion-recovery (FLAIR) revealed periventricular hyperintensities, which in addition to cerebral infarction, suggested multiple malignant lymphoma or HIV encephalopathy. Even with the initiation of anti-retroviral therapy (ART), his level of consciousness had gradually worsened along with acute gastrointestinal bleeding. One month after admission, the patient died due to pneumonia. The autopsy unexpectedly showed disseminated Kaposi sarcoma (KS), in addition to HIV encephalopathy and multiple central nervous system diffuse large B-cell lymphoma. KS lesions were found in the stomach, small intestine, liver, spleen, mesentery and lungs (Fig. 1). However, KS was not observed on his skin. Gross findings revealed multiple nodular lesions in the affected organs, and hematoxylin and eosin staining showed proliferation of spindle cells with vascular proliferation (Fig. 1). Immunostaining was positive for endothelial markers (CD31 and

* Corresponding author at: Laboratory of Clinical Immunology and Microbiology, National Institute of Allergy and Infectious Diseases, NIH, Bethesda, MD, 20852, USA.

E-mail address: hounamugun@gmail.com (H. Namkoong).

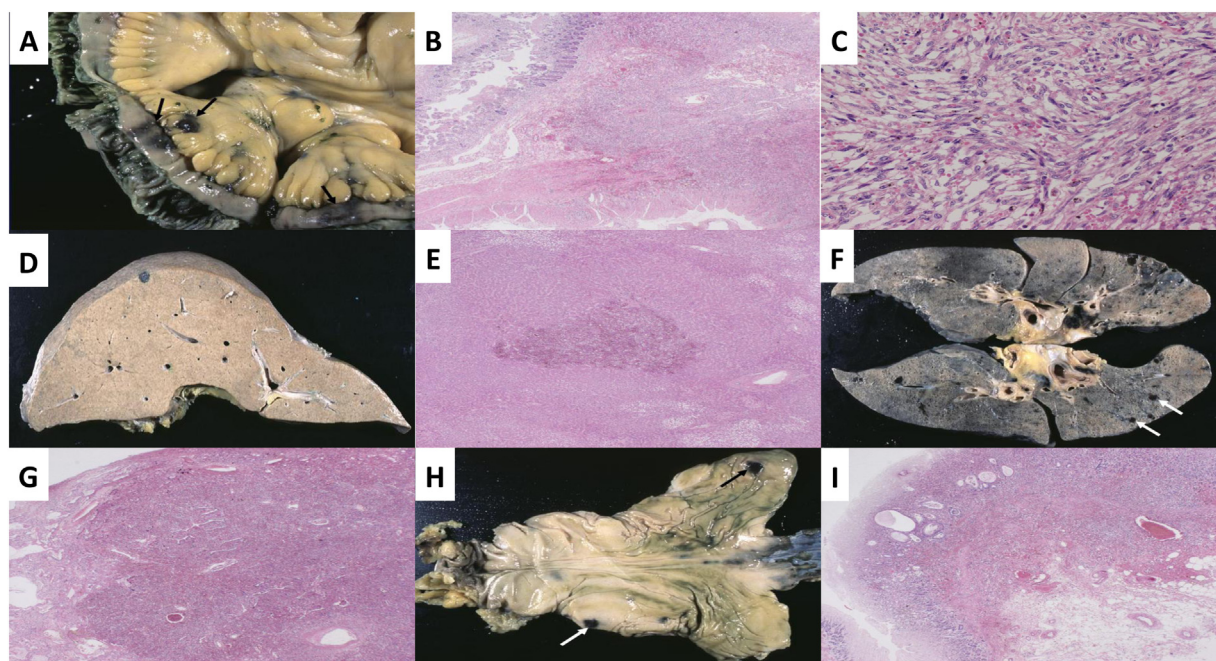


Fig. 1. A: Gross findings of the small intestine and mesentery showing nodular lesions (black arrow), B (low magnification) and C (high magnification): HE staining of the small intestine and mesentery showing nodular lesions from the submucosa to serosal layer, D: Gross findings of the liver showing diffuse nodular lesions (black arrow), E: HE staining of the liver showing proliferation of blood vessels and spindle cells (low magnification), F: Gross findings of the lung showing nodular lesions (white arrow), G: HE staining of the lung showing proliferation of spindle cells (low magnification), H: Gross findings of the stomach showing nodular lesions (white arrow), I: HE staining of the stomach showing proliferation of spindle cells in the stomach wall (low magnification).

von Willebrand factor) and lymphatic endothelial marker (D2-40), which are consistent with KS (Fig. 2).

KS is the most common tumor in patients with acquired immunodeficiency syndrome (AIDS), especially in MSM. KS is caused by the human herpes-virus 8 infection [1]. It has an indolent clinical course and mostly involves cutaneous lesions over the lower limbs,

trunk, and oral cavity [2]. In this case, autopsy revealed disseminated KS, which was unrecognized before his death. The gastrointestinal bleeding might be related to his gastrointestinal KS. We were unable to perform endoscopy for gastrointestinal bleeding due to his worsening clinical status. This case highlights the possibility of disseminated KS being present even in the absence of cutaneous lesions.

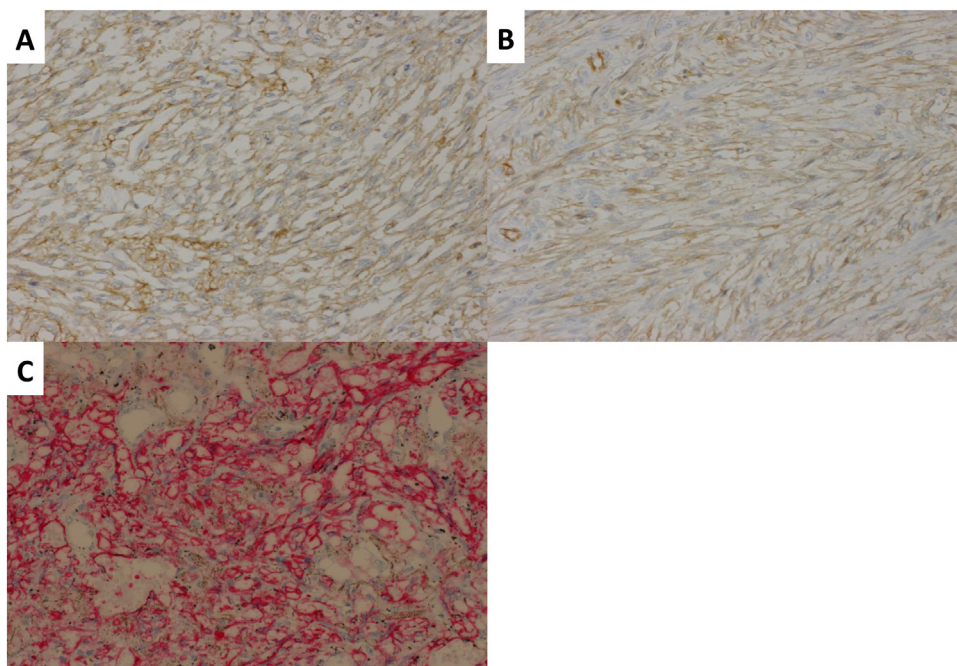


Fig. 2. A: Immuno-staining for von Willebrand factor showing positive findings, B: Immunostaining for CD31 showing positive findings, C: Immunostaining for D2-40 showing positive findings. HE: Hematoxylin and eosin.

Funding statement

The authors have no funding to report.

Informed consent

Written informed consent was unobtainable because the patient was deceased.

CRediT authorship contribution statement

Naoki Kawakami: Writing - original draft. **Ho Namkoong:** Conceptualization, Data curation, Project administration, Supervision, Writing - original draft, Writing - review & editing. **Masayuki Shimoda:** Data curation, Writing - review & editing. **Hiroshi**

Kotani: Data curation, Writing - review & editing. **Hiroshi Fujiwara:** Data curation, Writing - review & editing. **Naoki Hasegawa:** Supervision, Writing - review & editing.

Declaration of Competing Interests

The authors have declared that no competing interests exist.

References

- [1] Cesarman E, Damania B, Krown SE, Martin J, Bower M, Whitby D. Kaposi sarcoma. *Nat Rev Dis Primers* 2019;31(5):9, doi:<http://dx.doi.org/10.1038/s41572-019-0060-9>.
- [2] Etemad SA, Dewan AK. Kaposi sarcoma updates. *Dermatol Clin* 2019;37:505–17, doi:<http://dx.doi.org/10.1016/j.det.2019.05.008>.