










Human Immunodeficiency Virus-Associated Gastrointestinal Kaposi's Sarcoma: A Case Report

후천성 면역 결핍 바이러스와 연관된 위장관 카포시 육종: 증례 보고

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Kaposi's sarcoma (KS) is a multicentric human immunodeficiency virus-associated neoplasm characterized by multiple vascular nodules in the skin, mucous membranes, and viscera. Gastrointestinal acquired immunodeficiency syndrome (AIDS)-related KS is the most common visceral involvement reported in disseminated disease. Here, we present the findings of a rare case of KS involving multiple organs with abdominal pain and active bleeding in the colon. Multiple intraluminal lesions were found in the terminal ileum, sigmoid colon, and rectum by ileocolonoscopy, and in the jejunum and ileum by fluoroscopy. Abdominopelvic CT revealed multiple enhanced flat lesions in the ileum and enlarged lymph nodes. The diagnosis was confirmed by histopathology, and antiretroviral therapy was initiated as the treatment of choice for KS. Owing to the increasing number of AIDS patients, it is essential for radiologists and clinicians to be aware of the imaging characteristics of KS to protect physicians from indiscriminate exposure to AIDS.

Index terms Sarcoma, Kaposi; Acquired Immunodeficiency Syndrome; Small Intestine

INTRODUCTION

Gastrointestinal Kaposi's sarcoma (KS) is a rare acquired immunodeficiency syndrome (AIDS)-related malignancy. KS was first described by Moritz Kaposi in 1872 (1). Gastrointestinal KS can involve any level of the gastrointestinal tract with concomitant cutaneous compromise. The most common site is the duodenum (1). A red-purple sub-

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mucosal appearance in endoscopic and double-contrast barium studies can be observed in patients with gastrointestinal KS (2, 3). Fluoroscopic findings of gastrointestinal KS are rare; hence, to date, few cases have been documented. Herein, we report fluoroscopic findings of a case of KS involving the small bowel.

CASE REPORT

A 44-year-old man presented to our hospital with lower abdominal pain, especially after meals. He had no known disease, and his complete blood count, blood chemistry, and electrolyte profile were normal except for aspartate aminotransferase/alanine aminotransferase (51/59 U/L), erythrocyte sedimentation rate (42 mm/hr), and CD4 cell count (112/ μ L). The patient underwent esophagogastroscopy and ileocolonoscopy. The endoscopies revealed multiple reddish papular lesions in the stomach, terminal ileum, colon, and rectum (Fig. 1A-C). The mucosal lesions in the stomach and terminal ileum were biopsied. Pathology showed atypical spindle cell proliferation in the terminal ileum, which the pathologist suspected as KS. Additional immunohistochemistry was performed, which revealed that the biopsy sample was positive for CD34 and human herpesvirus 8 (HHV-8) and negative for actin. The patient was pathologically confirmed to have KS in the terminal ileum. The patient had never been

Fig. 1. A 44-year-old man with human immunodeficiency virus-associated gastrointestinal Kaposi's sarcoma, presenting with abdominal pain. **A-C.** Endoscopy shows multiple reddish papular lesions in the stomach (**A**) and terminal ileum (**B**). An ileocolonoscopy conducted after 4 months (**C**) reveals a reddish round elevated lesion in the sigmoid colon with focal active bleeding (arrows). **D.** Small bowel fluoroscopy shows multifocal polypoid lesions in the jejunum and ileum (arrows).

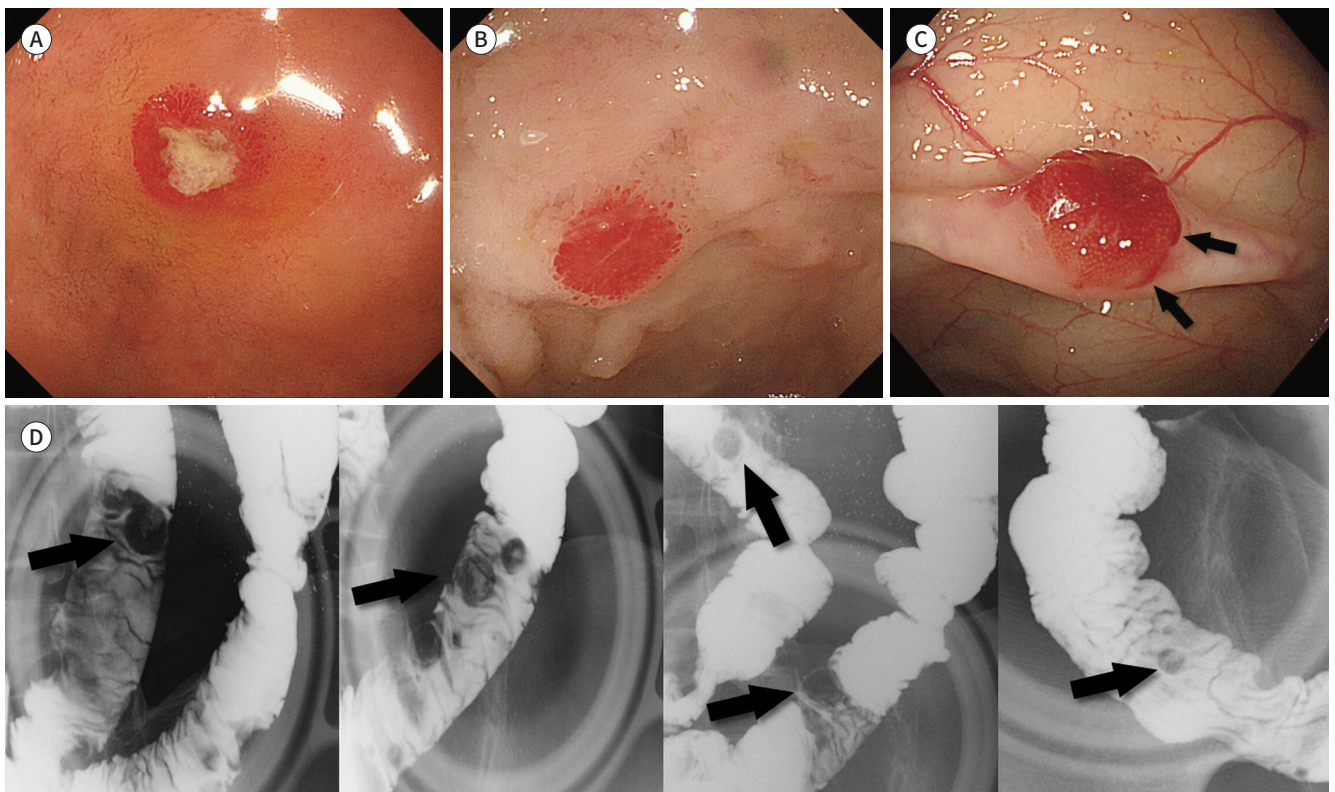
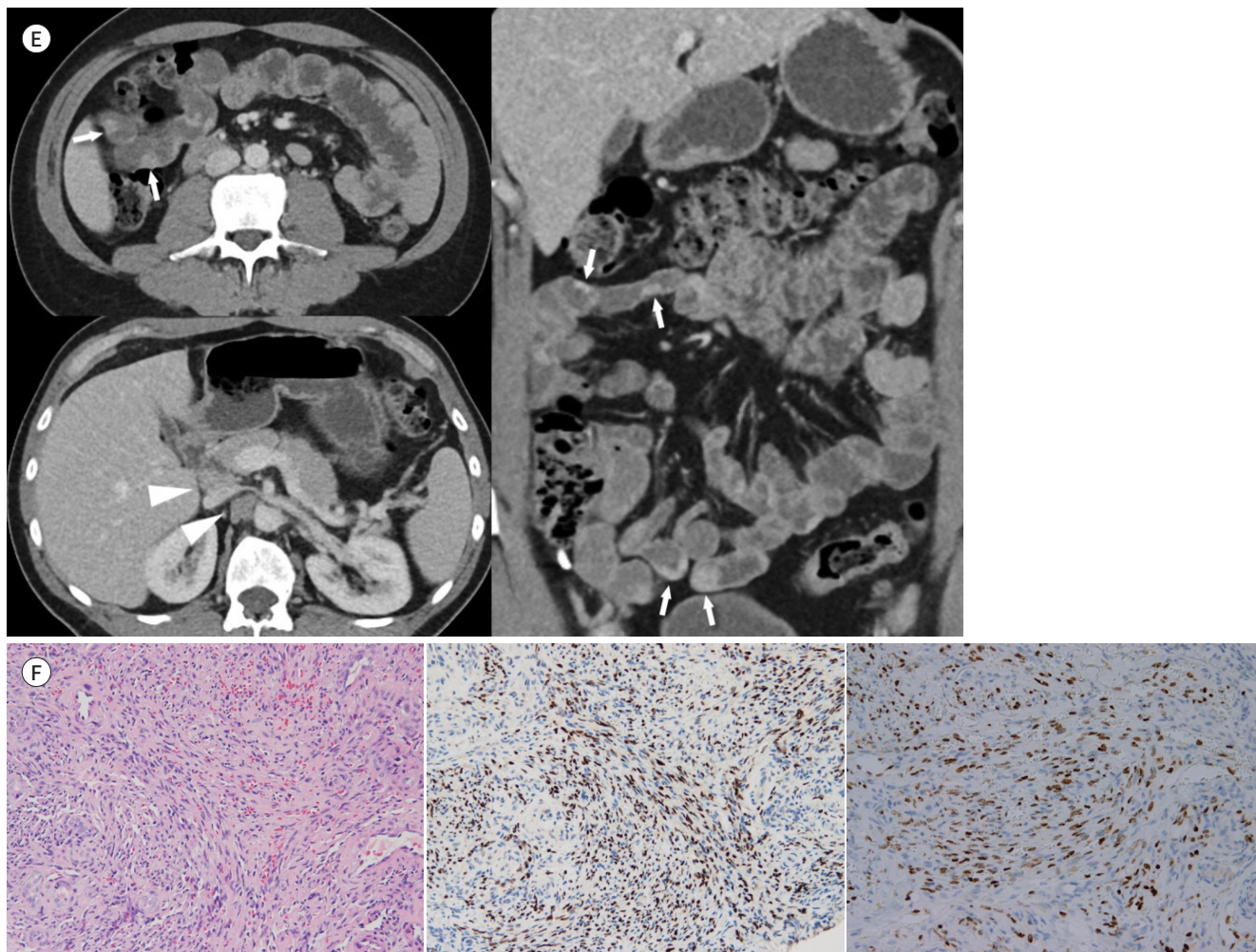


Fig. 1. A 44-year-old man with human immunodeficiency virus-associated gastrointestinal Kaposi's sarcoma, presenting with abdominal pain. **E.** Abdominopelvic CT images show multiple enhanced flat mucosal nodules (arrows) in the jejunum and ileum and enlarged lymph nodes at the portocaval and aortocaval areas (arrowheads).

F. Histological specimens ($\times 200$). Hematoxylin and eosin staining of the sarcoma shows slit-like spaces with spindle cell proliferation and red blood cell extravasation in the terminal ileum (left). Immunohistochemistry showing diffuse positivity for human herpesvirus 8 in spindle cells along the slit-like spaces in the terminal ileum (middle) and skin (right).



diagnosed with human immunodeficiency virus (HIV) infection. He confessed that he had undergone a skin biopsy at a local clinic about 6 months earlier, but the pathology was normal. He persistently had several purple skin papules, and described the lesions as gradually spreading throughout his body. After the biopsy, the skin lesions were also diagnosed as KS. Following pathologic confirmation of KS, further work-up was performed to classify the subtype of KS, which was finally confirmed as AIDS-related KS. The patient was initially treated with antiretroviral therapy. Four months later, the patient's hemoglobin was 6.1 g/dL, and he had hematochezia; subsequently, endoscopy was performed to detect active gastrointestinal bleeding. The endoscopy revealed multiple reddish round elevated lesions in the stomach, terminal ileum, colon, and rectum with focal active bleeding in the colon (Fig. 1C). The patient was treated with endoscopic hemostasis in the sigmoid colon and then received a blood transfusion. To assess the lesions in the jejunum and ileum, small bowel fluoroscopy was per-

formed, which revealed multifocal polypoid lesions in the distal jejunum as well as the distal and terminal ilea (Fig. 1D). Abdominopelvic CT was performed to evaluate the extent of the gastrointestinal KS. CT images revealed multiple enhanced flat mucosal nodular lesions in the ileum as well as enlarged lymph nodes at the portocaval and aortocaval area (Fig. 1E, F).

DISCUSSION

KS is a vascular tumor characterized by neoangiogenesis, proliferation of spindle-shaped cells, extravasated red blood cells, and vascular clefts and is related to HHV-8 infection; it is the most common neoplasm in HIV-infected patients, affecting 15–20% of this population (4). HHV-8 virus is known to be associated with KS in HIV seropositive patients. KS is one of the AIDS-related malignancies, such as high-grade B-cell non-Hodgkin lymphoma and invasive cervical cancer (5). KS is classified into four sub-types: epidemic or AIDS-associated, endemic, classic, and immunosuppression therapy-related. KS in the current case was classified as epidemic or AIDS-associated KS. Endemic KS is found in some populations in parts of Africa, and classic KS is found in older men of Mediterranean and Jewish lineage. The final type is immunosuppression-associated or transplantation-associated KS (2). In Korea, the cumulative number of patients with HIV increased in 2016 (6), and concerns of AIDS-related tumors are on the rise.

KS primarily manifests as skin lesions, lymph nodes, and the gastrointestinal tract, occurring in more than 15–35% of patients with AIDS-associated KS (1). However, the majority of cases are symptomless. A recent guideline indicated that CT scans and endoscopy are not warranted in the absence of symptoms (5). The overall incidence of gastrointestinal KS accounts for 25–50% of total KS cases but is underestimated because the patients are typically asymptomatic (7, 8).

Gastrointestinal KS should be differentiated from other common diseases that affect the gastrointestinal tract in patients with AIDS, such as cytomegalovirus infection, non-Hodgkin lymphomas, leiomyomas, rhabdomyosarcomas, high-grade pleomorphic sarcomas, or gastrointestinal stromal tumors. Multifocal involvement of the gastrointestinal tract, liver, spleen, and lymph nodes is a typical feature of KS in the abdomen, and the duodenum is the most commonly involved site. Gastrointestinal KS is diagnosed based on large masses and multiple nodules with or without central ulceration (“bullseye” or “target” lesions) observed in small bowel fluoroscopy and CT images that are not indicative of omental or peritoneal disease. On the other hand, AIDS-related lymphoma is characterized by a large or cavitory mass along the gastrointestinal tract, which can appear as peritoneal lymphomatosis (9).

Patients with gastrointestinal KS may rarely exhibit bleeding, obstruction, abdominal pain, vomiting, and diarrhea (7-9). In our patient, skin lesions, active bleeding, and abdominal pain were observed. Because active bleeding can be a life-threatening complication, it often requires endoscopic or angiographic diagnosis and treatment. We diagnosed gastrointestinal KS in this case with multiple modalities including CT, endoscopy, and fluoroscopy. The fluoroscopic and CT findings showed multifocal polypoid lesions in the distal jejunum and distal and terminal ileum. Small bowel fluoroscopy can help predict the extent of lesions, which is one of the prognostic factors of KS (10). With the increasing number of patients with AIDS, it

is essential for radiologists and clinicians to be aware of the imaging characteristics of KS and for physicians to accordingly take proper precautions and diagnose and treat HIV-infected patients. Treatment of gastrointestinal KS mainly involves palliative management and prevention of disease progression with highly active antiretroviral therapy (5). After initiation of highly active antiretroviral therapy, which is likely associated with improvement of CD4 cell counts, the incidence of this neoplasm decreased to less than 1% in patients infected with HIV.

In conclusion, we evaluated the multifocal gastrointestinal involvement of KS with active bleeding in this case by endoscopy, CT, and fluoroscopy. With the increasing number of patients with AIDS in Korea, radiologists and clinicians should be aware of the various imaging characteristics of KS.

Author Contributions

Conceptualization, P.S.H.; data curation, C.J.W.; formal analysis, L.H.J.; investigation, P.S.; supervision, P.S.H.; visualization, L.H.Y., C.D.H.; writing—original draft, L.H.J.; and writing—review & editing, C.S.J.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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후천성 면역 결핍 바이러스와 연관된 위장관 카포시 육종: 증례 보고

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카포시 육종은 후천성 면역 결핍 증후군(acquired immunodeficiency syndrome; 이하 AIDS)와 연관되어, 피부, 점막과 여러 장기를 침범하는 다발성의 혈관성 결절로 나타나는 신 생물을 일컫는다. AIDS와 연관되어 생기는 위장관 카포시 육종은 내장에 파종성 질환으로 가장 흔하게 발생한다. 우리는 다양한 장기를 침범하여 복통과 함께 장관 내 출혈을 유발한 드문 카포시 육종의 영상 소견에 대해 보고한다. 회장 대장내시경을 통해 말단 회장, S자 결 장, 직장 내 다양한 병변이 발견되었으며 소장 투시검사로 공장과 회장의 병변을 확인할 수 있었다. 복부 골반 전산화단층촬영에서 회장 내 조영증강된 다양한 납작한 병변과 복강 내 크기가 커진 림프절을 발견하였다. 조직병리학 검사에서 카포시 육종으로 최종 진단이 된 환 자는 항레트로바이러스제로 치료를 시행 받았다. 최근 AIDS 환자의 수가 늘고 있는 상황 에서, AIDS와 연관된 Kaposi's sarcoma의 영상학적 소견을 숙지하는 것은 영상의학과 의사를 비롯한 임상 의사들에게 빠른 진단과 치료뿐만이 아니라 예상치 못한 AIDS의 감염 위험에 노 출될 수 있는 의료진의 안전을 위해서도 필수적이다.

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