Kaposi's Sarcoma of the Small Intestine After Renal Transplantation: Radiological and Endoscopic Findings

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A case of Kaposi's sarcoma involving the small bowel two years after receiving a renal transplant is described. Immunosuppression had been achieved using cyclosporine A and prednisolone. Lesions extended from the duodenum to the ileum; radiologically, they were demonstrated on small bowel follow-through study and computed tomography as multiple small nodular intraluminal masses with or without central umbilication, and endoscopically, were seen as intramural mucosal elevations with a central crater-like ulceration.

Index Words: Intestinal neoplasms, CT Intestines, radiography

Kaposi's sarcoma is a rare malignant vascular tumor of unknown etiology, characteristically affecting the skin but sometimes involving other parts of the body. Patients with acquired immunodeficiency syndrome (AIDS) and AIDS-related disorders are predisposed to its development (1); it may also occur in organ transplant recipients who undergo prolonged or intensive immunosuppressive therapy (2-4).

The radiological manifestations of AIDS-related Kaposi's sarcoma of the intestinal tract have been infrequently demonstrated and reported (5-9), while the radiological findings of intestinal Kaposi's sarcoma after renal transplantation have not been previously reported. We report the occurrence of Kaposi's sarcoma of the small intestine, including radiological and endoscopic findings, in a patient who received renal transplantation.

Case Report

Two years prior to consultation, a 31-year-old female received a renal allograft because of chronic renal failure. Immunosuppression with cyclosporine A (main tenance dose: 200 mg daily) and prednisolone (30mg daily) was commenced; despite intermittent elevation of blood urea nitrogen and creatinine to levels of up to 27.5mg/dL and 2.1mg/dL respectively, her condition remained stable, but her hemoglobin value subsequently fell to 5.9g/dL. To determine the cause of this anemia, gastrofiberscopy was performed, and multiple small round to oval-shaped mucosal elevations with central ulcerations were seen in the duodenum and jejunum (Fig. 1). No definite mass was noted in the stomach. A biopsy specimen of the duodenum revealed Kaposi's sarcoma. A small bowel follow-through study showed multiple, small intraluminal nodular masses, predominantly involving the jejunum and responsible for distorting the mucosal pattern; they produced contour defects as well as intraluminal lucencies (Fig. 2). The nodules ranged in size up to 2cm. Abdominal computed tomography (CT) depicted multiple small nodular masses projecting into the lumen of bowel loops, including the duodenum, jejunum, and ileum (Fig. 3). At the time of diagnosis of Kaposi's sarcoma, there were no visible cutaneous and oral lesions; serologic tests for human immunodeficiency virus and cytomegalovirus were negative.
Discussion

Kaposi’s sarcoma was initially described in 1872 as a pigmented sarcoma of the skin with a tendency to involv the gastrointestinal tract. Histologically the tumor is characterized by irregular, vascular channels lined by endothelium, spindle-shaped cells, and varying degrees of mononuclear inflammatory infiltrates in the surrounding tissues. Until the AIDS epidemic, Kaposi’s sarcoma was considered a rare neoplasm, found mainly among elderly males living in Europe and the Mediterranean region, and following a slow, indolent clinical course in which skin lesions predominated. Interestingly, a more endemic and aggressive form was observed in Central Africa, starting around 1950. Kaposi’s sarcoma was also noted in renal transplant recipients from around 1970 (7, 10).

Kaposi’s sarcoma accounts for only 0.02% to 0.07% of all malignancies in general populations (3). Nowadays, however, the epidemiology of the disease has changed significantly because of its strong association with AIDS. Patients with AIDS and AIDS-related disorders are predisposed to its development. Another predisposing factor for the occurrence of Kaposi’s sarcoma is an organ transplantation in relation with immunosuppressive therapy; it is generally agreed that there is an increased risk of malignant disease in immunosuppressed patients following transplantation. Inability of the immune system to combat neoplastic cellular activity leads to increased incidence of neoplasia in this patient population. Some immunosuppressive agents such as azathioprine, cyclophosphamide, and cyclosporine may directly damage DNA and cause cancer. An epidemiologic study showed a 400- to 500-fold increase in the incidence of Kaposi’s sarcoma in renal transplant recipients, compared with controls of the same ethnic origin (3, 4). According to Suleiman et al (2), the incidence of kaposi’s sarcoma in patients with renal transplantation was 3.4% and the mean duration from

Fig. 1. Endoscopic findings of Kaposi’s sarcoma involving the duodenum. There are multiple small round to oval shaped raised lesions with umbilication. The lesions are intensely red as compared with the normal duodenal mucosa.

Fig. 2. Small bowel follow-through findings of Kaposi’s sarcoma (A, B). Overhead films of the small bowel show multiple small intramural nodules that project into the lumen, thereby producing filling defects (arrow heads). Some of the lesions are coalescent together.
transplantation to diagnosis was 13.6 months (range, 4 months to 4 years). Penn (3) reported that Kaposi’s sarcoma appeared at an average of 21 months (range, 2 to 225.5 months) after transplantation, including that of the heart and kidney. In our patient, Kaposi’s sarcoma developed 2 years after renal transplantation. The incidence of Kaposi’s sarcoma in renal transplants at our hospital has been estimated as roughly 1%.

If the lesions are flat, radiologic studies usually fail to show Kaposi’s sarcoma, but elevated lesions may appear as intraluminal filling defects (discrete sharp submucosal nodules 6 mm to 3 cm in size) of variable size and number; central umbilication may be seen, and there is normal intervening mucosa without ulceration. Contiguous nodules are sometimes so numerous that they resemble thumbprinting or irregular fold thickening (6–8). Our case showed typical polyloid or nodular lesions, or raised lesions with and without umbilications on small bowel follow-through study. CT appearances of Kaposi’s sarcoma most likely reflect the size of the nodular lesions. In this case, therefore, not all the intraluminal nodular masses were visualized on CT image, as was the case with the small bowel follow-through study. CT or magnetic resonance imaging may be helpful in showing the extent of involvement including retroperitoneal and mesenteric lymphadenopathy (7, 9). Endoscopically, the lesions are discrete, flat or raised, and intensely red because of the underlying histopathology; that is, a capillary hemangiosarcoma with endothelial proliferation; endoscopy is more sensitive than radiologic examinations in depicting superficial incipient lesions represented by small macular mucosal discolorations (7).

Non-Hodgkin’s lymphoma may, as in the case of Kaposi’s sarcoma, also occur in the small intestine of AIDS or transplantation patients and may not be radiologically distinguishable; metastatic melanoma or multiple polyps may have similar appearances. The diagnosis depends on the biopsy findings (7, 9, 10).

Organ transplantation has been performed in many institutions in Korea; radiologic findings of organ transplantation-induced malignancies therefore need to be carefully evaluated. In conclusion, the described radiographic findings are, in addition to the presence of intramural polyloid lesions in the gastrointestinal tract, indications of Kaposi’s sarcoma if present in renal transplantation with immunosuppressive therapy, and particularly when associated with hemorrhagic skin lesions.

References

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Fig. 3. Axial CT scan at the level of the grafted kidney reveals a small round intraluminal polyloid mass on the wall of ileum which is well contrasted by the ingested oral contrast medium (arrows).
신장이식수술 후 발생한 소장의 Kaposi 육종: 방사선학적 및 내시경적 소견

신장이식수술을 받은 다음 2년 후에 발생한 소장의 Kaposi 육종에 대해 보고한다. 신장이식 후 사용된 면역억제제는 cyclosporine A와 prednisolone이었다. Kaposi 육종은 심장 저장에서부터 회장까지 분포하였다. 이 병변은 저장장성 심장장병벽, 소장조영술 및 전산화단층촬영영상에서 소장내벽에 다수의 작은 결점성 종괴로 관찰되었으며, 일부 종괴에서는 종괴의 중앙부위가 함몰되어 보였다. 내시경상 병변은 중앙부위에 분화구 같은 케양을 갖는 장내 점막이 용기된 종괴의 소견을 보였다.