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CLINICAL CASE REPORT

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Abdominal Kaposiform Hemangioendothelioma Associated With Lymphangiomatosis Involving Mesentery and Ileum

A Case Report of MRI, CT, and 18F-FDG PET/CT Findings

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Abstract: Kaposiform hemangioendothelioma (KH) is a rare vascular tumor of intermediate malignancy that occurs mainly in the childhood. Adult patients with KH are rare. Imaging findings of KH have rarely been reported before. We present magnetic resonance imaging (MRI), computed tomography (CT), and fluorine-18-fluorodeoxyglucose (^{18}F-FDG) positron emission tomography (PET)/CT findings in an adult patient with KH associated with lymphangiomatosis involving mesentery and ileum.

A 22-year-old female complained of a 9-month history of intermittent melena, weakness, and palpitation. Laboratory tests revealed anemia and hypoproteinemia. Fecal occult blood test was positive. Abdominal enhanced MRI and CT showed a large abdominal mass involving mesentery and ileum. On enhanced MRI, there were many hypervascular nodules in the mass. On FDG PET/CT, the mass and the nodules showed slight FDG uptake. Small bowel capsule endoscopy showed numerous grape-shaped red nodules in the luminal wall of the involved ileum. The patient underwent resection of the abdominal mass and a segment of the ileum invaded by the abdominal mass. KH arising within lymphangiomatosis involving mesentery and ileum was confirmed by pathology. After surgery, the patient's symptoms improved.

This is the first case of KH associated with lymphangiomatosis involving mesentery and ileum. In this case, the lymphangiomatosis overshadowed the small tumor nodules resulting in unusual imaging findings. Familiarity with these imaging findings is helpful for diagnosis and differential diagnosis of KH.

(*Medicine* 95(6):e2806)

Abbreviations: ^{18}F-FDG = fluorine-18-fluorodeoxyglucose, CT = computed tomography, KH = kaposiform hemangioendothelioma, MRI = magnetic resonance imaging, PET = positron emission tomography.

Editor: Pradeep K. Mittal.
Received: November 25, 2015; revised: January 15, 2016; accepted: January 20, 2016.
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AD and LZ contributed equally to the study.
The authors have no conflicts of interest to disclose.

ISSN: 0025-7974
DOI: 10.1097/MD.0000000000002806

INTRODUCTION

Kaposiform hemangioendothelioma (KH) is a rare vascular tumor of intermediate malignancy that occurs mainly in the childhood and is often associated with Kasabach–Merritt phenomenon or lymphangiomatosis or both.[1–4] Histopathologically, KHs consist of irregular, infiltrating nodules of compressed vessels with resemblance to Kaposi sarcoma.[1] About 82% of KHs occur in patients within the first decade of life and 45% within the first year.[1] Adult patients with KH are rare. Most of KHs present as a superficial or deep extremity mass.[1] Occasionally, KHs may involve chest and abdominal wall, retroperitoneum, mediastinum, scrotum, tongue, spleen, lung, heart, liver, and stamoch.[1–9] Multifocal KH in multiple visceral organs have been reported.[7,8] Complete surgical excision is the most effective treatment of KH.[1] Approximately 42% to 58% of patients present with Kasabach–Merritt phenomenon marked by severe thrombocytopenia and a variable degree of anemia[1,2] and two-thirds of KHs exhibit lymphatic abnormalities.[1] Imaging findings of KH have rarely been reported before.[10–12] In this paper, we present magnetic resonance imaging (MRI), computed tomography (CT), and fluorine-18-fluorodeoxyglucose (^{18}F-FDG) positron emission tomography (PET)/CT findings in an adult patient with KH associated with lymphangiomatosis involving mesentery and ileum.

CASE REPORT

A 22-year-old female was admitted to our hospital because of a 9-month history of intermittent melena, weakness, and palpitation. She had undergone treatment for anemia, but had no significant improvement. On clinical examination, the patient presented with pale skin and a tender abdomen. Her past medical history was unremarkable. Laboratory tests revealed anemia and hypoproteinemia. Fecal occult blood test was positive. Abdominal T2-weighted MR images showed a large hyperintense mass in the middle and lower abdomen (Figure 1A). This mass showed hypointensity on unenhanced T1-weighted MR images. Enhanced T1-weighted MR images showed many hypervascular nodules scattered in the mass (Figure 1B and C). Abdominal unenhanced CT showed this mass was hypodense involving mesentery and ileum. There were multiple relatively hyperdense nodules in the mass. The mass and the nodules showed slight enhancement on enhanced CT (Figure 1D). For further evaluation of the patient, ^{18}F-FDG FDG PET/CT was performed showing slightly increased FDG uptake of the mass and the small nodules (Figure 2). Mesenteric panniculitis was suspected. Small bowel capsule endoscopy showed grape-shaped red nodules in the luminal wall of the ileum (Figure 3A).

The patient underwent resection of the abdominal mass and a segment of the ileum (length: 60 cm) invaded by the

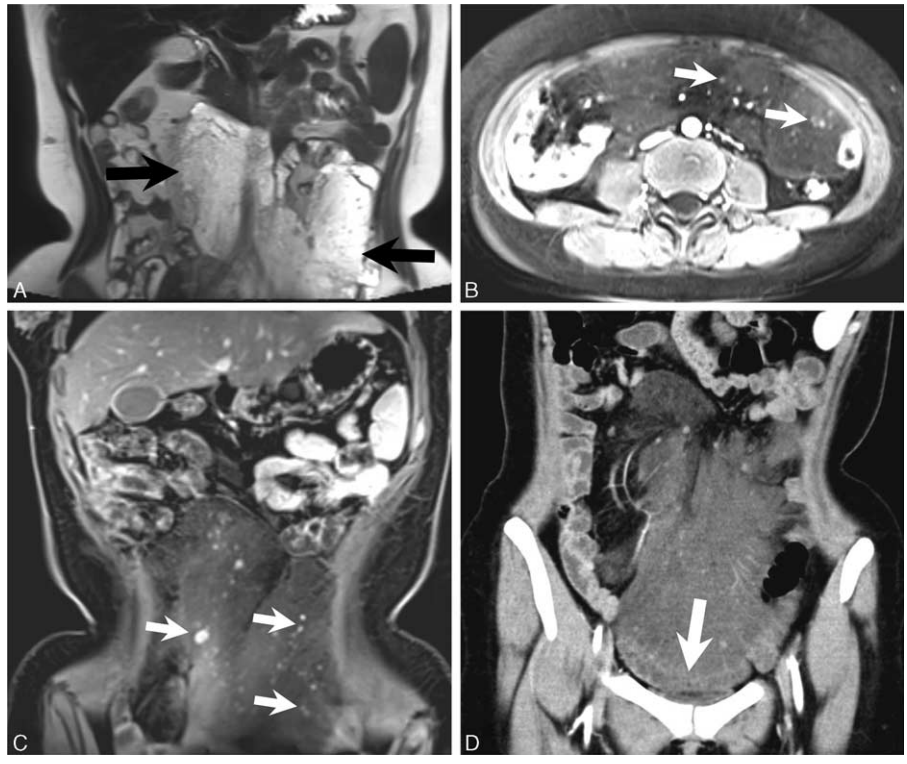

FIGURE 1. Abdominal coronal T2-weighted MR image (A) showed a large hyperintense mass (arrows) in the middle and lower abdomen. Transverse (B) and coronal (C) enhanced T1-weighted MR images showed many hypervascular nodules (arrows) scattered in the mass. Coronal enhanced CT (D) showed the mass involved mesentery and ileum (arrow).

abdominal mass. Photomicrograph of the resected specimen showed proliferation and dilation of the mucosal lymphatics in the ileum, containing a large amount of red blood cells (Figure 3B). In the abdominal mass, there are many irregular tumor nodules (Figure 3C). Scattered throughout the tumor nodules were epithelioid or glomeruloid areas with abundant capillary sized vessels of attenuated lumina. There was dense hyaline sclerosis in the tumor nodules. The tumor cells were positive for CD31, CD34, and D2-40. Ki-67 staining showed the proportion of the positive tumor cells was about 1%. Marked lymphatic proliferation containing red blood cells encircled the tumor nodules (Figure 3D). The imaging and histopathologic findings were consistent with KH associated with lymphangiomatosis involving mesentery and ileum. After surgery, the patient's symptoms improved. Follow-up laboratory tests showed normal red blood cell count and hemoglobin level.

DISCUSSION

KHs occur mainly in children and rarely in adults. About 82% of these tumors occur within the first decade of life and 45% within the first year.[1] The most common locations are

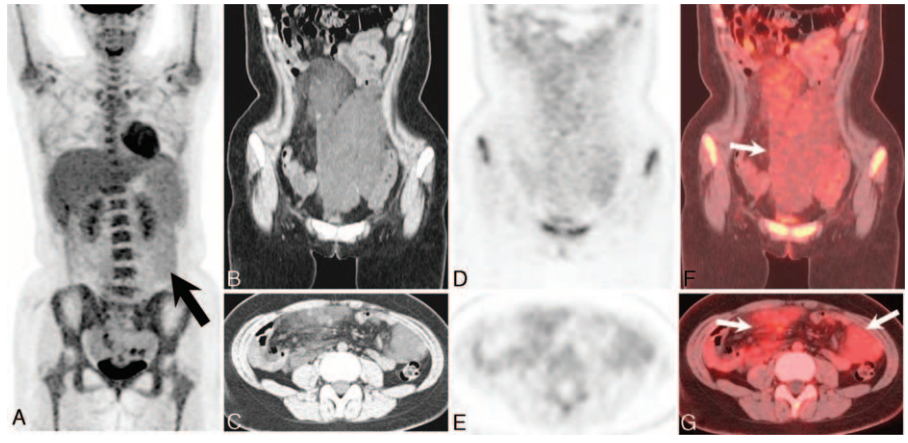

FIGURE 2. Maximum intensity projection PET (A), coronal (B) and transverse (C) CT, corresponding PET (D and E), and fused (F and G) images showed slightly increased FDG uptake of the mass (arrows). Mesenteric panniculitis was suspected.

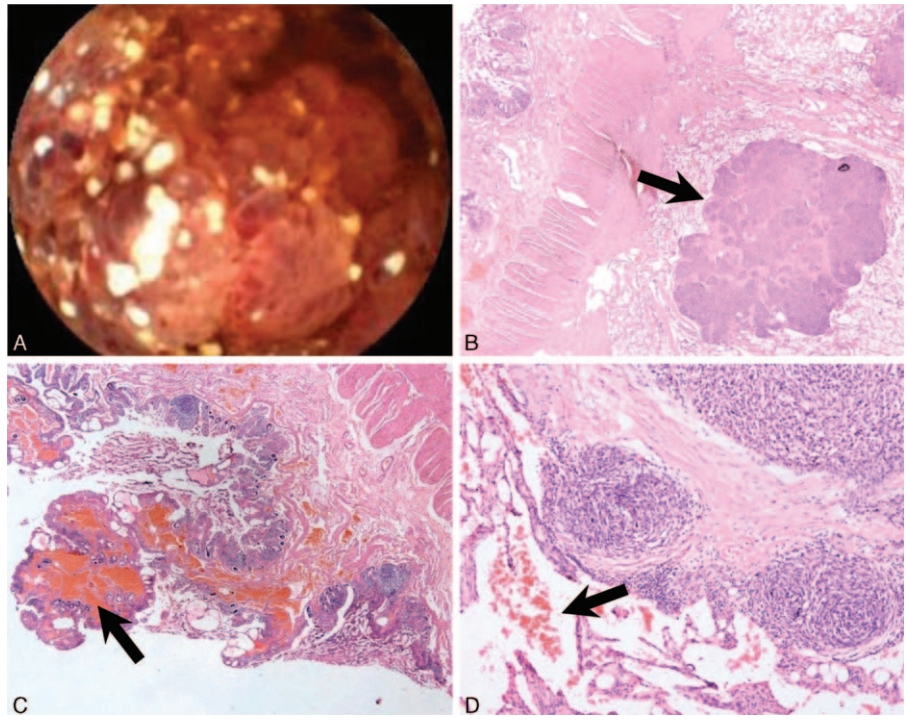

FIGURE 3. Small bowel capsule endoscopy showed grape-shaped red nodules in the luminal wall of the ileum (A). Photomicrograph (C, hematoxylin and eosin, original magnification ×20) showed proliferation and dilation of the mucosal lymphatics of the ileum, containing a large amount of red blood cells. In the abdominal mass, there are many irregular tumor nodules (arrow) (B, hematoxylin and eosin, original magnification ×20). Marked lymphatic proliferation containing red blood cells (arrow) encircled the tumor nodules (D, hematoxylin and eosin, original magnification ×100).

superficial or deep soft tissues of the extremities followed by head and neck.[1] Approximately half of the KH patients can develop Kasabach–Merritt phenomenon marked by severe thrombocytopenia and a variable degree of anemia.[1,2] Patients with Kasabach–Merritt phenomenon tended to be younger.[1] Why Kasabach–Merritt phenomenon develops in the setting of KH is unclear. One explanation is that unique architectural and/or endothelial differences in KH that may promote consumption coagulopathy.[1] In KH, small convoluted capillaries arise directly from large vessels in a serial or linear fashion, arguably creating a situation that results in turbulence leading to platelet activation and aggregation.[1] This patient only had anemia and no thrombocytopenia. Numerous grape-shaped red nodules in the luminal wall of the ileum revealed by the small bowel capsule endoscopy were dilated lymphatics containing red blood cells. Intestinal bleeding and subsequent anemia may result from the ruptured lymphatics. Where was the blood of the dilated lymphatics came from was unclear. It may be from the tumor vessels. Further investigation is needed. Approximately two-thirds of KHs exhibit lymphatic abnormalities.[1] Like this case, the lymphatic proliferation can result in a mass lesion which supporting for the idea that lymphatic abnormalities are not simply the result of lymphatic obstruction with lymphangiectasis but represent an intrinsic part of the lesion.[1] One hypothesis is that KH begins as a lymphatic malformation onto which a vascular component is engrafted.[1] Another hypothesis is that KH actually produces lymphatic endothelial growth factors, which incite proliferation of adjacent lymphatics.[1,2]

CT and MRI findings of KH associated with lymphangiomatosis have rarely been reported before.[8–12] On CT or MRI, KHs tend to demonstrate ill-defined margins and involvement of multiple tissue planes. KHs show hypointensity similar to that of muscle on T1-weighted images, heterogeneously hyperintense relative to that of muscle on T2-weighted images and heterogeneous enhancement.[12] In this case, the lymphangiomatosis overshadowed the small tumor nodules resulting in unusual imaging findings. Enhanced CT clearly showed the border of the abdominal lymphangiomatosis and the involved organs, but unable showed the small tumor nodules in the lymphangiomatosis. Compared with enhanced CT, enhanced MRI clearly detected the hypervascular tumor nodules on the lower signal background of the lymphangiomatosis. The lymphangiomatosis was better demonstrated on T2-weighted images with remarkable hyperintensity.

We searched the electronic database of PubMed with terms of ''fluorine-18-fluorodeoxyglucose,'' ''positron emission tomography,'' and ''kaposiform hemangioendothelioma.'' To our knowledge, 18F-FDG PET/CT findings of KH have not been reported before. In this case, the tumor nodules showed slight FDG uptake mimicking benign lymph nodes. The lymphangiomatosis also showed mild FDG uptake, which may be due to accumulation of FDG in the lymphatics. This case indicates, although rare, KH associated with lymphangiomatosis should be considered in the differential diagnosis of abnormal mesenteric FDG accumulation, including lymphangioma,[13] mesenteric panniculitis,[14–16] and mesenteric malignancies with mesenteric panniculitis.[16]

In conclusion, this is a rare case of KH associated with lymphangiomatosis involving mesentery and ileum. This case indicates finding hypervascular nodules on the background of lymphangiomatosis may be helpful for diagnosis of KH.

ETHICAL REVIEW AND CONSENT

Ethical approval was obtained from the Ethics Committee of Changhai Hospital, Shanghai, China. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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