Protein-loss into retroperitoneal lymphangioma:
Demonstration by lymphoscintigraphy and blood-pool scintigraphy
with Tc-99m-human serum albumin

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A rare, benign congenital lymphangioma has been reported to occur frequently in the neck and axilla, but rarely in the retroperitoneal space. We report a case of a retroperitoneal lymphangioma associated with hypoproteinemia caused by protein-loss into the tumor. In this case, lymphoscintigraphy with subcutaneously injected Tc-99m-human serum albumin (HSA) disclosed the communication between the tumor and the lymphatic system, and sequential abdominal scintigraphy with intravenously injected Tc-99m-HSA revealed the protein loss into the tumor. Abdominal scintigraphy with Tc-99m-HSA injected intravenously or subcutaneously is occasionally useful for determining the etiology of hypoproteinemia.

Key words: retroperitoneal lymphangioma, Tc-99m-HSA, lymphoscintigraphy, hypoproteinemia

CASE REPORT

A 14-year-old Japanese boy, who had a one-month history of bloody and watery diarrhea, was admitted to our hospital. The symptoms were more obvious in the morning and exacerbated by exercise. On physical examination, low blood pressure (systolic/diastolic, 90/50 mmHg), anemic palpebral conjunctiva, slight abdominal distention and pitting edema in the anterior lower legs were noted. No abdominal mass was palpable. Laboratory tests revealed anemia and hypoproteinemia; red blood cell count, $371 \times 10^4/\mu l$; hemoglobin concentration, 9.9 g/dl; hematocrit, 32.3%; serum total protein and albumin concentrations, 3.9 g/dl and 1.96 g/dl, respectively.

Abdominal ultrasonography revealed a large cavernous or cystic mass, and ascites. Abdominal computed tomography (CT) showed a well-circumscribed cystic mass spreading over the whole abdomen. The inside of the mass showed water density on the CT (Fig. 1). The border between the mass and pancreas was indistinct, and a part of the mass seemed to infiltrate into the bowel wall. After an injection of contrast material, normal abdominal vasculature was delineated but no abnormal enhancement was demonstrated in the mass. The bowel was compressed by the mass. In particular, jejunum and ileum were displaced to the anterior space of the abdomen.

Since the borders of the mass with the pancreas and the colon were unclear on the CT, endoscopic retrograde cholangiopancreatography (ERCP) and barium enema were performed. On ERCP, there was no obvious abnormal finding. On barium enema, multiple submucosal tumors were suspected in the left transverse to the sigmoidal colon. The lesions ranged from 3 mm to 15 mm in diameter.

Angiography showed no obvious anatomical abnormality, but demonstrated stretched vasculature around the splenic flexure, descending colon and pancreas, indicating the presence of a hypovascular tumor extending to these organs. No apparent vascular proliferation was observed in the tumor.

Based on these findings, a diagnosis of lymphangioma was strongly suspected. To investigate the relationship between the tumor and the lymphatic system, lymphoscintigraphy with Tc-99m-human serum albumin (Tc-99m-HSA) was performed. Seventy-four MBq of Tc-99m-HSA was injected subcutaneously into the webbing...
Fig. 1 Plain abdominal CT at about the level of the first (A) and of the third lumber vertebra (B) demonstrate a well-circumscribed cystic mass of water density extending over the whole abdomen (arrow heads). The bowel is compressed to the right lower side by the mass.

Fig. 2 Anterior and posterior whole body images obtained 30 minutes (A) and 3 hours (B) after the subcutaneous injection of Tc-99m HSA. On the images at 30 minutes, there is no apparent abnormal activity in the region of the tumor. The abnormal activity extending from mid to left abdomen, which corresponds to the cystic tumor on CT, becomes apparent on the images at 3 hours after the injection (arrow heads).

between the first and second toes, and whole body imaging was done 30 minutes, one hour, two and three hours after the injection. The lymphoscintigrams showed abnormal activity, which increased with time, in the abdomen (Fig. 2). The activity was localized to the region of the tumor. This finding proved the communication between the tumor and the lymphatic system, an important clue for diagnosing lymphangioma.

To clarify the etiology of the hypoproteinemia in this patient, blood pool scintigraphy with Tc-99m-diethylene triamine pentaacetic acid-human serum albumin (Tc-99m-DTPA-HSA) was performed. Seven hundred and forty MBq of Tc-99m-DTPA-HSA was intravenously injected as a bolus, and sequential images of the abdomen were taken 2.5, 5.5 and 24 hours after the injection. The obtained images showed abnormal activity increasing gradually with time. The location of the abnormal activity was not in the bowel, but in the tumor as seen on the lymphoscintigraphy (Fig. 3). This finding indicated that protein loss did not occur into the bowel, but into the tumor.

Based on these findings, a diagnosis of protein loss into the retroperitoneal lymphangioma was made. The patient underwent surgery for sclerotherapy of the tumor with OK-432 as a sclerosing agent. The abdominal surgery revealed a multilocular cystic mass spreading over