

CASE REPORT

Treatment of Retroperitoneal Cavernous Lymphangioma: A Case Report

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Abstract A 32-year-old man who complained of recurrent nausea and vomiting was admitted to our hospital. The contrast-enhanced computed tomography revealed a cystic mass located behind the duodenum which was suggestive of lymphangioma. Laparoscopic resection of the retroperitoneal mass was successfully performed. The postoperatively pathological examination confirmed the diagnosis of cavernous lymphangioma. Ultrasound and enhanced CT can be used for making a preoperative diagnosis. Once symptoms of the disease develop, complete surgical resection should be performed.

LYMPHANGIOMA is a rare benign cystic lesion arising from the lymphatic system. The common types of lymphangiomas include cavernous lymphangiomas, cystic lymphangiomas and simple lymphangiomas.^[1] Usually, most patients with lymphangioma are asymptomatic. When the lump compresses the adjoining structures, clinical symptoms will appear. This lesion mostly occurs in children, and is seldom found in the retroperitoneum.^[2, 3] Here we report a case of a young man with a cavernous lymphangioma located in the retroperitoneum, who presented with recurrent vomiting, and was successfully treated with laparoscopic excision of the lesion.

CASE DESCRIPTION

A 32-year-old man presented with recurrent nau-

sea and vomiting for one month without abdominal pain, constipation, or fever. He lost three kilograms in the previous month. Furthermore he has Down syndrome identified at birth. Physical examination was normal and routine blood test was within normal limits. The abdominal ultrasound showed an irregular-shaped cystic space-occupying lesion which had multiple compartments and a clear boundary involving the right retroperitoneum (**Figure 1**). The contrast-enhanced computed tomography of the abdomen revealed an irregular low-density (18 HU) lesion of 7.0 cm×4.6 cm in size with some focal spot-like calcification in it, located behind the second part of the duodenum (**Figure 2**). Based on the above findings, a clinical diagnosis of retroperitoneal lymphangioma was made. Considering this situation that the cystic lesion was too adjacent to the right ureter to identify the ureter easily during surgical excision of the lymphangioma, a stent was planted in the right ureter one day prior to surgical removal of the tumor.

Laparoscopic resection of the retroperitoneal



Figure 1. Abdominal ultrasound showing an irregular cystic lesion in the right retroperitoneal space (arrow).



Figure 2. Contrast-enhanced computed tomography of the abdomen revealing a hypodense lesion in the retroperitoneum (arrow), which displaced the duodenum anteriorly, and was close to the inferior vena cava and the right renal hilum concurrently.

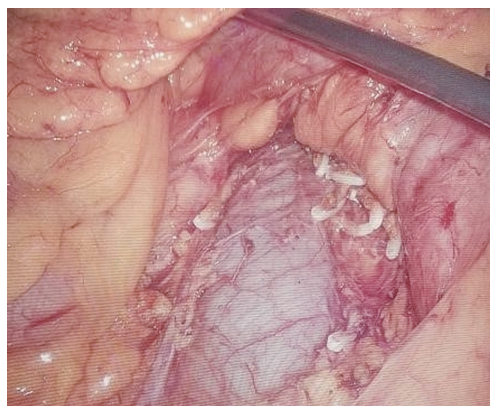


Figure 3. The intraoperative image showing the tumor bed formed by the inferior vena cava.

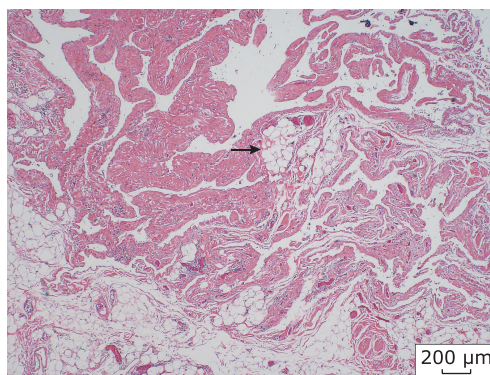


Figure 4. The microscopic appearance of the lymphangioma showing a mass that comprises of lymphangitic sac (arrow) of varying sizes, and contains a small number of smooth muscle bundles and lymphocytes. HE staining

mass was successfully performed, in which the mass was completely removed and no lymphatic fluid leaked out. It took 94 minutes and blood loss was 10 ml. Intraoperatively, we found that it was apparent that the mass arose from lymphatic vessels which were located behind the duodenum (**Figure 3**). The postoperative recovery was uneventful with a hospital stay length of six days. The postoperative histology confirmed the diagnosis of cavernous lymphangioma (**Figures 4**). At the last follow-up visit of six months, he was symptom-free.

DISCUSSION

Approximately 95% of lymphangiomas generally occur in the neck or axilla of children. Lymphangiomas originating from the lymphatic system of the retroperitoneum is quite rare. Since abundant loose connective tissues are distributed in the retroperitoneal space,

patients of retroperitoneal lymphangiomas are usually asymptomatic until the mass grows large enough to displace or compress the adjacent organs, for example when duodenum is involved, the patient will develop nausea and vomiting.

At present, both ultrasound and CT are considered the most valuable imaging modalities for retroperitoneal lymphangioma diagnosis.^[4] On ultrasound, the lesion appears as hypoechoic cystic mass, with or without some areas of calcification. When cavernous lymphangioma is accompanied by cavernous hemangioma, red and blue blood flow signals can be found in parts of the lumen.^[5] On CT, the lesion appears as multiple tortuous and dilated cystic structures, which often have a honeycomb appearance, with a clear boundary, hypodense contents, and a visible enhancement of the capsule on contrast injection. Calcification of the capsule wall can occur, but is not common.^[6, 7] Ultrasound and CT have been

applied in the differential diagnosis of retroperitoneal lymphangioma from intestinal duplication cyst, retroperitoneal teratoma, mesenteric cyst, etc. Intestinal duplication cyst that is a rare congenital malformation in which a thick cyst wall enfolds fluid, debris, or blood components, shows a typical “three-layer” appearance, representing the echogenic mucosa, hypoechoic muscle layer and echogenic serous membrane of intestinal walls on ultrasound.^[7] Retroperitoneal teratoma is a type of germ cell tumor which contains skin, hair, bone, and several different types of cells like those existed in various organs and glands, which are visible on CT image. Mesenteric cysts is a relatively rare disease that could be attributed to migration of intestinal cells or diverticulum into mesentery, with a characteristic pathologic change of a cystic wall lacking a muscular layer and a nerve plexus. Therefore, its typical CT finding is a thin cystic wall filled with fluid. On ultrasound, this disorder appears as a hypoechoic cystic mass with an occasional thin septum.^[8] The representative CT finding of retroperitoneal lymphangioma is an irregular shaped, small and low density cystic lesion located in the retroperitoneum, which anteriorly displaces or compresses, but not infiltrates or destroys the surrounding tissues, with no enhancement in the cystic wall and no mural nodules.

As a benign tumor, surgical resection of retroperitoneal lymphangioma is required only for symptomatic cases.^[9] Great care is needed when separating nerves and blood vessels from the mass. For this case, no recurrence was found after six months of follow-up. However, more studies are required to determine recurrence of lymphangioma.^[9]

In conclusion, ultrasound and enhanced CT can be used to make a preoperative diagnosis of retroperitoneal lymphangioma. Once the patients present with clinical symptoms, complete surgical resection should be performed.

Conflict of Interests Statement

The authors declare no conflict of interests.

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