lymphatic malformations are a form of congenital vascular malformations that have the characteristic of involving the head and neck. Abdominal lymphatic malformations are very rare, representing 5% of all lymphatic malformations.\textsuperscript{1} They may be found in the abdominal cavity, mesenterium, retroperitoneum, solid organs (spleen, liver and pancreas) and gastrointestinal system. Clinical findings often vary by the size and localization of the lesion. Of the patients, 88% manifest symptoms during childhood.\textsuperscript{2} In adulthood, they are frequently noticed incidentally by abdominal imaging procedures in individuals who are otherwise asymptomatic. When symptomatic, patients may have abdominal pain due to organomegaly or pressure effects, distention, nausea, vomiting, constipation, diarrhea and palpable mass in the abdomen.\textsuperscript{3,4} Gastrointestinal bleeding and protein-losing enteropathy have also been reported in rare cases involving the mesenterium and gastrointestinal system.\textsuperscript{6,8} Lymphatic malformations manifest most frequently as mesenteric lymphatic malformations in the abdomen.\textsuperscript{9} Retroperitoneal lymphatic malformations are very rare in this group and represent less than 1% of all abdominal lymphatic malformations.\textsuperscript{10} Here, we present a case involving a large-sized lymphangiomatosis localized in the retroperitoneum in a patient who presented to our clinic with epigastric discomfort and was diagnosed with serous cystic tumor with endoscopic ultrasonography before.

\textbf{Keywords:} Abdominal cystic lymphangioma; cystadenoma; peritoneum
CASE REPORT

Written informed consent obtained from the patient.

The 38-year-old female patient presented to the outpatient clinic with abdominal distension. The patient had no other accompanying symptoms such as abdominal pain, weight loss, nausea or vomiting. Her history did not involve trauma or intra-abdominal surgery and her familial history also involved no relevant findings. Patient’s results from the physical examination, laboratory tests, tumor marker analysis and upper gastrointestinal system endoscopy were within normal ranges. Abdominal ultrasound (USG) demonstrated a 4-cm cystic lesion extending along the pancreas and therefore endoscopic ultrasound (EUS) was requested. The result of the performed endoscopic USG was reported as lesion which could be consistent with serous cyst adenoma with diffuse septation along the retropancreatic space (Figure 1). Because the patient had no symptoms at that time, fine-needle aspiration was not performed. The result of her abdominal magnetic resonance imaging (MRI) performed to investigate diffusiveness of the lesion was reported to involve a cystic lesion in the retroperitoneal area with retropancreatic localization, consistent with lymphangiomatosis extending along the left retrocrural region up to the diaphragm (Figure 2, Figure 3). The patient is still asymptomatic and 1-year follow-up MRI has the same features.

DISCUSSION

Lymphangioma is a congenital clinical phenomenon which can occur in any organ except brain that lacks lymphatics. More than 90% of the patients are individuals below 2 years of age and it is often seen in the head and neck. It is frequently asymptomatic in adulthood. Lymphangiomatosis is a subtype of lymphangioma and it is the cystic dilation of the lymphatic channels, and is very rare, and can either be limited with an organ or tissue (spleen, liver, thoracic cavity etc.) or may be more generalized.
Abdominal lymphatic malformations are one of the clinical phenomena that we keep in mind when we come across intra-abdominal cystic lesions. Patients may present with involvement that is restricted to the mesenterium, retroperitoneum, liver, spleen, renal, pancreas or with more diffuse involvement, although rarely. Retroperitoneal lymphatic malformations are very rare in this group and represent less than 1% of all abdominal lymphatic malformations. It should be remembered that malformations that are adjacent to the pancreas as in our case may be confused with pancreatic cystic tumors due their cystic structural features. Contrast MRI or computed tomography (CT) and EUS sampling may be helpful in differential diagnosis. CT or MRI is critical in such cases to investigate multisystemic involvement and lesion diffusiveness. Clinical findings depend on the site of involvement of the lesion. Follow up is sufficient in asymptomatic patients, while sclerotherapy, percutaneous or laparoscopic aspiration or cyst marsupialization in local or regional involvements may be attempted in patients who are symptomatic and have diffuse cystic lesions. Patients who are not suitable for these and those who are refractory and symptomatic are candidates for surgery. Intestinal lymphangiectasia usually respond to dietary changes such as a following low-fat diet and substitution of long-chain fatty acids with mid-chain fatty acids. Octreotide and tranexamic acid may be used in patients with moderate gastrointestinal bleeding.

In conclusion; lymphangiomatosis may present with diffuse organ involvement or may be large in size at a single site. Cases that were confused with metastatic disease due to diffuse bone involvement, hematologic malignancy due to spleen involvement and infectious causes (echinococcus, kala-azar etc.) and kidney tumor due to perirenal involvement have been reported. Adjacency to the pancreas in the retroperitoneal area lead us to suspect serous cystic adenoma of the pancreas. An experienced radiologist seems to be diagnostically critical at this point. It is evident that pathologic sampling is required for undifferentiated patients.

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