Disseminated lymphangiomatosis presenting as chylous ascites and diagnosed with endoscopic ultrasound

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A 43-year-old male patient presented with progressive abdominal distension for one month. Ascitic tap yielded milky fluid, and the fluid triglycerides were 711 mg/dl consistent with chylous ascites. Cytology of fluid done thrice revealed no malignant cells. Contrast-enhanced computed tomography showed cystic lesions in the pancreas, liver, spleen, the lesser sac and gross ascites [Figures 1 and 2]. Fine-needle aspiration (FNA) from the cystic lesion of the liver yielded serous fluid, and cytological examination was inconclusive. Flourodeoxyglucose (FDG)-positron emission tomography did not show any FDG-avid lesion. Endoscopic ultrasound (EUS) revealed enlarged mediastinal lymph nodes with cystic spaces along with the splenic, hepatic, lesser sac and pancreatic cysts [Figure 3a and b]. EUS guided FNA from the pancreatic, as well as lesser sac cyst, [Figure 4] yielded chylous fluid with normal carcinoembryonic antigen and amylase and increased triglycerides. Cytological examination of the FNA from pancreatic and lesser sac cysts as well as subcarinal lymph node revealed lymphocytes and macrophages in a fluid background suggestive of lymphangioma [Figure 5]. The patient was started on high protein and medium chain triglyceride diet. At 3 months follow-up the patient is better, and ascites has resolved.

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Lymphangiomas are rare benign cystic malformations of the lymphatic system commonly seen in children and adults are rarely affected.[1] They commonly occur in the head and neck region and very rarely they are widespread, and this condition is called as "lymphangiomatosis."[2] Lymphangiomatosis may involve any organ except brain and spinal cord as these organs lack lymphatic system.[2,3] Histologically, these are endothelium lined cystic spaces that contain chyle.[1,4] Abdominal lymphangiomatosis is an uncommon clinical entity which must be suspected in the presence of multiple cystic lesions in various abdominal organs.[4] The abdominal lymphangiomatosis may present with abdominal pain, gastrointestinal bleeding, protein losing enteropathy or may be asymptomatic.[2] They may rarely present with chylous ascites because of the peritoneal rupture of the cystic lymphangiomas.[5]

On imaging, these closely mimic other causes of cystic lesions and therefore the correct diagnosis preoperatively is difficult.[2-4] EUS better characterizes these cystic lesions and has been useful in making the diagnosis of lymphangiomas.[3] Although various morphological features of lymphangiomas have been described in EUS but none of them is characteristic. However, EUS guided FNA may help in achieving correct diagnosis. Aspiration of chylous fluid that has elevated triglyceride content with numerous

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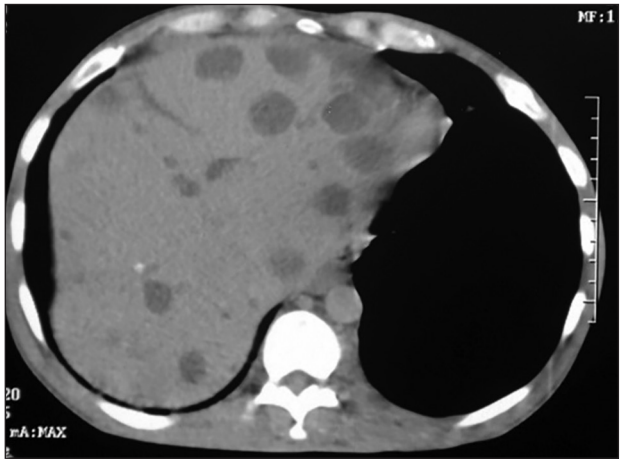

Figure 1. Multiple cystic lesions in liver

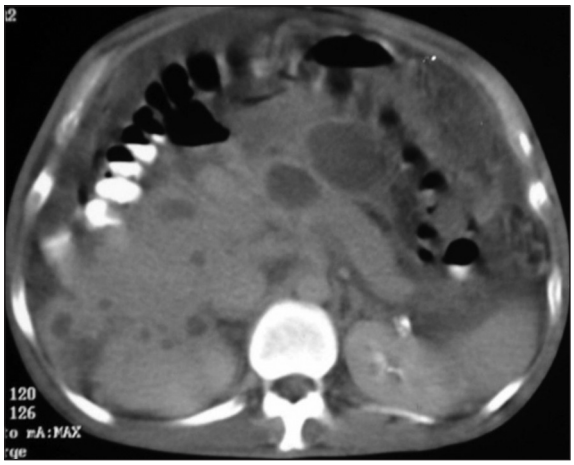

Figure 2. Pancreatic and lesser sac cysts

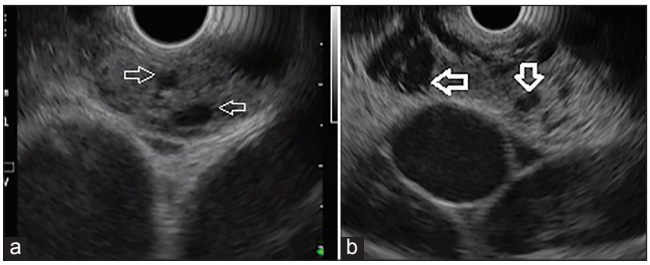

Figure 3. (a and b) Mediastinal lymph nodes with cystic spaces

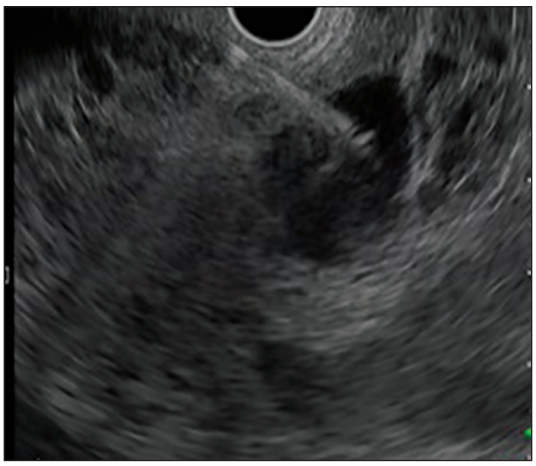

Figure 4. Endoscopic ultrasound guided aspiration of lesser sac cyst

lymphocytes on cytological examination may suggest a diagnosis of lymphangioma as was in our case.[3,6]

The EUS which was done for evaluation of the cystic lesion in the pancreas in this case revealed cystic lymph nodes. The presence of cystic lymph-nodes is unique and may also be seen with malignant metastasis from various primaries, but cystic lymphangioma represents an uncommon cause of cystic lymph nodes.[7] All in all, cystic lymphangiomatosis is an uncommon clinical entity in the adulthood and must be considered as a possibility in patients with multiple organs involved with cystic lesions.

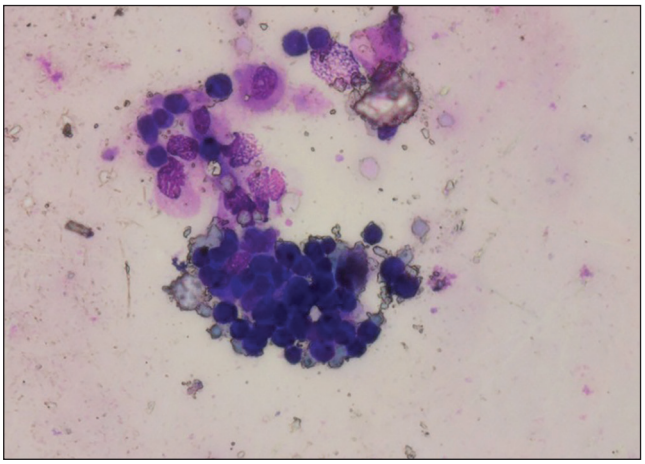

Figure 5. Microphotograph of fluid sediment showing collection of lymphocytes and few macrophages in a fluid background (May–Grunwald–Giemsa stain, original magnification ×400)

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