Cystic lymphangioma of the adrenal gland: Report of a case and review of the literature
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1. Introduction
Cystic lymphangiomas are rare benign lesions of the lymphatic vessels. These tumors originate from the lymphatic endothelial cells and are thought to be due to ectasia or abnormal development of lymphatic vessels. Most of the time these tumors appear in the neck or axilla, and intra-abdominal lymphangiomas only account for 5% of all lesions. The rarity of this disease and the lack of report render the diagnosis and management of this entity challenging. We report a case of left adrenal cystic lymphangioma in a female patient and review the current literature.

2. Case presentation
- 38-year-old woman known for anxiety disorder
- Constant epigastric pain, multiple episodes of vomiting
- CT: cystic-like retroperitoneal mass on the left side measuring 13.4 x 7.2 x 5.2 cm (Fig. 1)
- CT-guided puncture of the cyst: cytology negative for malignant cells and compatible with cystic lymphangioma
- MRI: lesion compatible with a cystic lymphangioma originating from the left adrenal gland (Fig. 2)
- Surgical resection of the cystic lesion: left en-bloc adrenalectomy in laparotomy (Fig. 3)
- Pathology: ovoid, cystic mass measuring 8.5 x 4.3 x 2.8 cm. Cystic space lined by a single layer of flattened cells, with occasional pseudopapillae formation and bands of smooth muscle in the wall (H&E stain). Strong immunoreactivity for D2-40, PROX1, CD31, and bands of smooth muscle in the wall (H&E stain). Strong immunoreactivity for D2-40, PROX1, CD31, and absence of staining for CD34 and CKAЕ1/АЕ3 (Fig. 4).
- Diagnosis of cystic lymphangioma originating from the left adrenal gland was then confirmed.
- Postoperative course: uneventful, patient discharged on day 5
- 12-month follow-up: no more symptoms

Figures 1-4 are not available in the text, but they are mentioned in the context of the case presentation.

3. Discussion
To our knowledge no more than 30 cases of adrenal cystic lymphangioma have been described in the literature. Table 1 summarizes the cases reported in the English-written literature since 2000. As confirmed in our case report, age at symptom onset usually ranges from 30 to 50 years. Cystic lymphangioma can occur in both adrenals, but more often the right side is affected. It also occurs more frequently in women.

Of note, lymphangiomas have an endothelial origin. The most probable etiology is a developmental abnormality or ectasia of the lymphatic vessels.

Differential diagnosis of a retroperitoneal cystic-like lesion includes primary adrenal tumors, metastatic adenocarcinoma, angiomyomas, pseudocysts, multicytic mesotheliomas, or adrenal cysts. Immunohistochemistry is an important tool to help differentiating this pathological entity from other diagnosis. Lymphangiomas usually display D2-40, CD31, and PROX1 positivity. Most of the time, these tumors are non-secretory and are discovered incidentally during a radiological exam or a surgery. Symptomatic tumors can induce pain, fever, gastrointestinal disturbances, or hypotension. Asymptomatic incidentally-discovered cystic lymphangioma can just be followed up as there is no risk of malignant degeneration.

In conclusion, cystic lymphangioma of the adrenal gland is a rare pathology that should be included in the differential diagnosis of cystic lesions of the adrenals. However, the diagnosis of this pathology can be difficult and challenging. MRI seems to be a good diagnostic modality to detect degeneration or intracystic hemorrhage. If the patient is symptomatic, definitive treatment is surgery.

4. References