Adrenal lymphangioma: a rare cystic lesion of the adrenal

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SOURIAL MW, VAN ROSSUM N, SABBAGH R. Adrenal lymphangioma: a rare cystic lesion of the adrenal. *Can J Urol* 2013;20(5):6968-6970.

We herein report the case of a left adrenal lymphangioma in a 52-year-old asymptomatic female and review the pertinent literature relatable to this rare, benign lesion.

Key Words: adrenal, lymphangioma, laparoscopy

Introduction

Adrenal lymphangioma (AL) are rare, benign lesions that are usually discovered incidentally during imaging for unrelated complaints. Cystic adrenal lesions are rare, and few reports of their appearances are found in the literature. We report the case of a cystic AL in a 52-year-old female and review the pertinent literature relatable to this benign lesion.

Case report

We report the case of a 52-year-old female referred for an indolent, slow growing, asymptomatic left adrenal mass. This mass was incidentally discovered in 1995 during a work up for total abdominal hysterectomy which she underwent for abnormal uterine bleeding. At that time, a computed tomography (CT) scan showed a 1.5 cm x 2.5 cm non-calcified, non-enhancing left adrenal nodule consistent with adenoma. Fifteen years later, a follow up CT scan showed a two fold increase, the mass now measuring 3.3 cm x 5.8 cm, still non-enhancing with minimal peripheral calcification and a density of 16 Hounsfield units, consistent with "probable adenoma." A year later, the lesion was stable in size measuring 3.4 cm x 5.1 cm, however some internal septa with thick calcification and more peripheral calcifications appeared, Figure 1. The proposed diagnosis was an adrenal cyst with dystrophic calcification but it was atypical with a significant growth within 16 years. An endocrinologic work up including serum electrolytes, plasma renin-aldosterone, dexamethasone suppression test, androgens (DHEAS, androstenedione, testosterone, LH/FSH), and urinary catecholamines and metanephrines, was normal failing to show a functional adrenal mass. An uneventful laparoscopic left adrenalectomy was performed.

Final pathology report revealed an 8 cm x 4 cm x 3 cm left adrenalectomy specimen with an embedded 4 cm cystic lesion with smooth yellowish contours

Accepted for publication July 2013

Acknowledgment
We'd like to thank Patrice Tessier, pathology resident, for the pathology figures.

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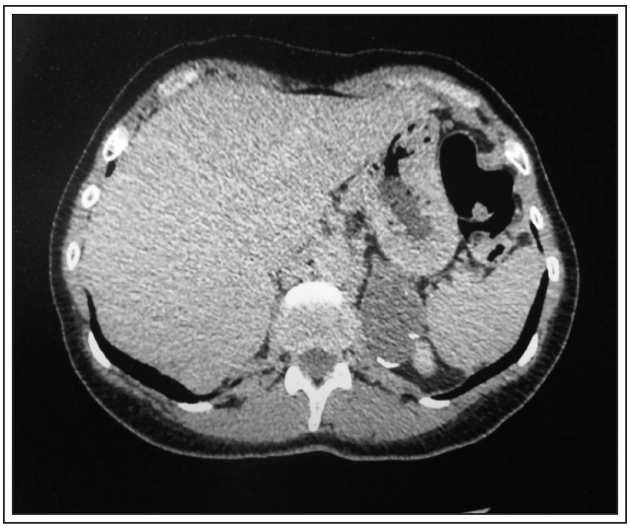

Figure 1. Non-contrast CT showing the hypodense 3.4 cm x 5.1 cm left adrenal lesion with internal septa with thick calcification in addition to the peripheral calcifications.

and few intraluminal calcifications. Microscopic analysis showed a multilocular cyst whose walls were lined with endothelial cells, Figure 2. Some of the loculations contained an amorphous eosinophilic substance that resembled lymph, without presence of red blood cells, Figure 3. Immunohistochemical analysis showed positivity of the cystic wall cells to CD31 and CD34 staining, and negative AE1/AE3 keratin staining, consistent with the diagnosis of an adrenal lymphangioma.

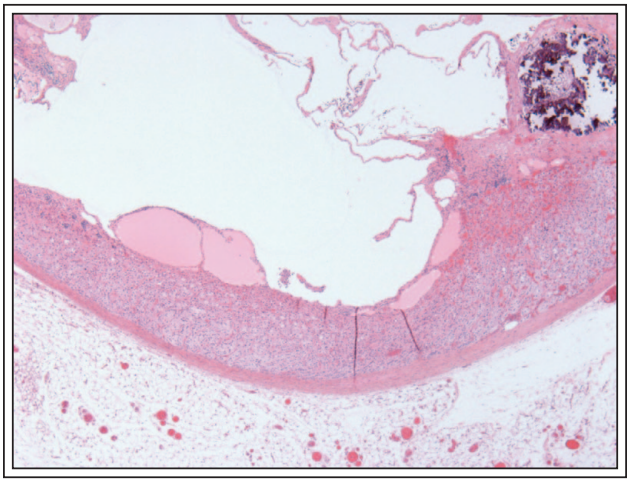

Figure 2. Adrenal gland (dark pink, middle) compressed by multiloculated cystic lesion (top). Calcification in the top right corner (4x).

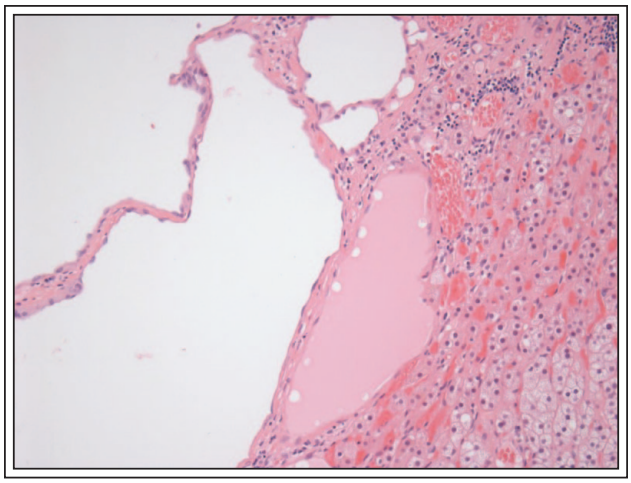

Figure 3. Cystic cavities lined with flat, non-atypical, endothelial cells. Note the absence of red blood cells in the cystic structures (200x).

Discussion

There are four types of adrenal cysts which are classified based on pathological findings: pseudocysts, parasitic cysts, epithelial lined cysts, and endothelial lined cysts. Endothelial lined cysts are further divided into lymphangiomatous or angiomatous cysts.[1] Adrenal lymphangiomas (AL), also known as cystic AL, are exceedingly rare, benign lesions that are usually asymptomatic and discovered incidentally. Cystic AL were initially discovered at autopsy, but advancements in radiographic imaging have led to earlier clinical detection of these lesions.

CT scan is classically more useful than ultrasonography to define adrenal lesions and assess for contrast enhancement. Adrenal cysts are fairly uncommon lesions and may commonly be mistaken for adenomas. Unlike adenomas, cystic lesions may be associated with malignancy (metastases, cystic adrenal carcinoma, pheochromocytoma) in as much as 7% in one reported series.[2] Rozenblit et al[3] concluded that the CT finding of a nonenhancing, thin-walled (≤ 3 mm) mass with or without wall calcification allows differentiation of an adrenal cyst from an adenoma. Calcification has been noted in 15% to 70% of adrenal cysts[4] and are typically peripheral but may also be nodular or intra-luminal. Because of the known risk of associated malignancy in adrenal cysts, surgical resection remains the standard of care in patients with a normal-appearing contralateral adrenal gland. Small, asymptomatic, thin-walled lesions maybe safely observed with active surveillance.[3]

Lymphangiomas are benign, cystic malformations of the lymphatic vessels, more frequently found in childhood with 90% discovered below 2 years of age.[5] They are located typically in the neck, axillary, or mediastinal regions. Abdominal lymphangiomas account for approximately 5% of all lymphangiomas, and these are typically located in the mesentery of the small intestine, omentum, mesocolon, or retroperitoneum. Adrenal lymphangiomas are exceedingly rare, and only about 50 cases have been reported in the literature, with Ellis et al[6] reporting the largest known series with nine AL reported over a period of 24 years. They commonly occur in the third to sixth decade of life, with a female, right-sided predominance. AL are typically asymptomatic, non-secreting lesions that are discovered incidentally on imaging for unrelated causes. If they are symptomatic, they may present with non-specific abdominal pain,[7] back pain[8] or as a palpable abdominal mass.[9]

Immunohistochemical staining includes CD31 and CD34 which are vascular markers that label both blood and lymphatic endothelial cells. D2-40 is a monoclonal antibody which is directed against podoplanin, a transmembrane protein expressed in lymphatic endothelial cells,[10] which makes it more specific as a lymphatic tumor marker. AE1 and AE3 are cytokeratin markers that further confirm the lymphatic rather than the mesothelial nature of the cystic lining.[6]

In conclusion, AL are exceedingly rare, benign, cystic lesions of lymphatic origin which are commonly asymptomatic and discovered incidentally. These cystic lesions are difficult to accurately characterize on radiographic imaging, and diagnosis is ultimately made on microscopic and immunohistochemical studies. We present the case of a cystic AL discovered incidentally in a 52-year-old female. □

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