

Cystic Adrenal Lymphangioma — Report of two cases and review of the literature

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Abstract

Cystic adrenal lymphangiomas are very rare, benign vascular lesions. They are usually found during a work up for abdominal pain or incidentally during imaging studies for an unrelated cause.

We report two cases of cystic adrenal lymphangiomas. They presented with flank discomfort, hypertension and flushing. Their laboratory findings were in normal limits. Radiologic imagings showed adrenal cystic neoplasm and the patients underwent adrenalectomy. Histopathologic examination and immunohistochemical findings were consistent with lymphangioma.

Cystic lymphangiomas may imitate other adrenal

neoplasms and must be kept in mind in the clinical and radiologic differential diagnosis of cystic adrenal lesions.

Keywords: Adrenal gland, Adrenal cyst, Lymphangioma.

Introduction

Lymphangiomas are benign vascular lesions and commonly located in the neck, axillary region and mediastinum. Adrenal lymphangiomas, also known as cystic adrenal lymphangiomas, are very rare and most often found incidentally during abdominal imaging studies or abdominal surgery or autopsy. There are less than 50 cases reported in the literature.¹⁻⁵ However, this entity may be seen more frequently with improvements in imaging techniques. Despite the advancements in radiographic techniques,

adrenocortical neoplasms and pheochromocytomas remain in clinical and radiologic differential diagnosis of these lesions. Recently we encountered two cases of cystic adrenal lymphangiomas. A purpose of this paper is to describe pathological, radiological and clinical features of our two cases and review the literature.

Case-1:

A 56-year-old man was admitted to our hospital with a history of hypertension and flushing lasting for two months. Physical examination and laboratory findings were normal. Abdominal ultrasonography revealed a 7 cm, lobulated cystic mass with septae located in the right adrenal gland. Magnetic resonance imaging showed a 7X5X4.5 cm cystic lesion revealing low signal intensity on

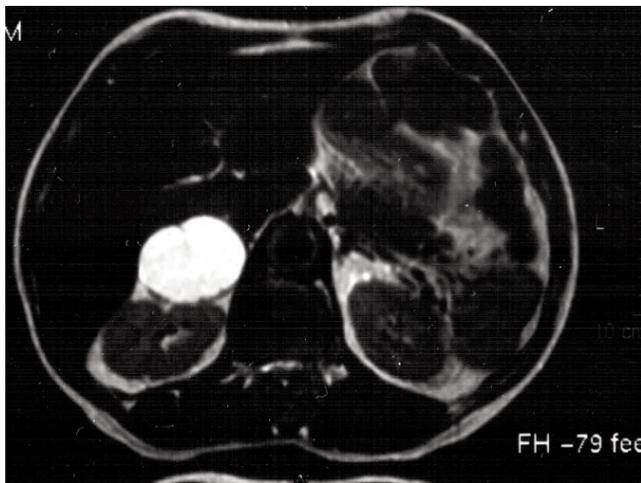


Figure-1: Magnetic resonance imaging of case 1 showed a 7X5X4.5 cm cystic lesion.

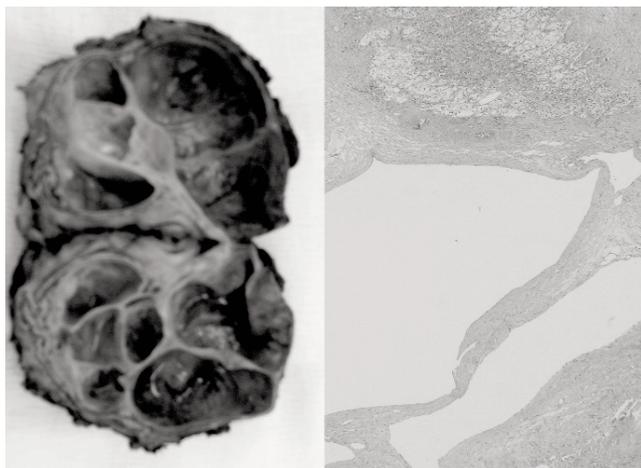


Figure-2: Macroscopic examination of case 1 showed, multi-septae cystic lesion surrounded by a rim of yellow orange adrenal gland and histologic sections showed multiple irregularly shaped cystic spaces lined by flat, bland and simple cells adjacent to the normal-appearing adrenal cortex (H&E, X40).

T1A weighted images and high signal intensity on T2A weighted images (Figure-1). Right adrenalectomy was performed for definite diagnosis. Macroscopically, adrenalectomy specimen weighed 123 gr and measured 8X7X5 cm. Cut sections revealed a 7X6X6 cm multi-septae cystic lesion filled with clear serous fluid and surrounded by a rim of yellow orange adrenal gland measured 3.5X1.5X0.6 cm (Figure-2).

Case-2:

A 46-year-old man was admitted to our hospital with left flank discomfort. On physical examination he had tenderness to palpation on left flank. Laboratory findings were normal. Abdominal ultrasonography displayed a 3 cm focal hypodense lesion at the upper pole of the left kidney. Magnetic resonance imaging showed a 3.5X3X2 cm cystic lesion revealing low signal intensity on T1 weighted images and high signal intensity on T2 weighted images. The patient underwent left adrenalectomy with a clinical and radiological diagnosis suspecting a pheochromocytoma. Grossly the adrenal gland measured 7X4X2 cm with a 2.5X1.5X1 cm multilocular cystic lesion.

Pathologic and Immunohistochemical Findings:

Microscopically, the lesions showed multiple irregular shaped cystic spaces lined by flat, bland and simple cells adjacent to the normal-appearing adrenal cortex (Figure-2). The cells lining the spaces had no atypia or mitosis. Immunohistochemically these cells stained positive with CD31 and CD34 and negative with pancytokeratin, confirming their endothelial origin. Case 1 revealed calcification foci in the cyst wall and septae. There was no evidence of associated adrenal hyperplasia, adenoma, carcinoma or pheochromocytoma in both of the cases. Postoperatively, the patients recovered without complications.

Discussion

Primary adrenal cysts are very rare lesions and often detected incidentally during radiologic investigation or at autopsy. The frequency of adrenal cysts seen at autopsy studies range from 0.064% to 0.18%.¹⁻⁶ Histologically, adrenal cysts have been classified into four groups: 1- Pseudocysts (39%) 2-Parasitic cysts (7%) 3- Epithelial cysts (9%) 4- Endothelial cysts (45%), the latter are subdivided into angiomatous and lymphangiomatous cysts.²⁻⁵ Adrenal cystic lymphangiomas are rare benign vascular lesions and are thought to arise from the abnormal development and/or ectasia of lymphatic vessels or from blockage of proximal lymphatics or trauma.²

Lymphangiomas typically occur in children but adrenal lymphangiomas occur at all ages with a peak

incidence between the third and sixth decades.¹⁻⁶ Recently Ellis et al. reported 9 cases of adrenal cystic lymphangioma, 6 female and 3 male patients with a mean age of 42 years. None of their cases was seen in paediatric age group. All of their lesions were unilateral with an average size of 4.9 cm (range 2.0cm- 13.5cm).² The presented cases were males but their ages and tumour sizes were consistent with the literature findings.

The differential diagnosis of adrenal cysts includes primary or metastatic, benign or malignant cystic neoplasms, haemorrhage secondary to trauma, bleeding disorder, burns, shock or toxemia, and infections like echinococcal cyst.^{2,5} On ultrasound, the diagnosis of cystic lymphangioma is suggested by the presence of a well-marginated, anechoic lesion. If calcification or internal debris is present, ultrasound may show more complicated appearance with internal echoes.¹⁻⁶ Computed tomography demonstrates, nonenhancing hypodense lesion. MRI features of adrenal lymphangioma are similar to those of other lymphangiomas that arise elsewhere in the body. On MRI, uncomplicated adrenal cysts are low in signal intensity on T1-weighted images and high on T2-weighted images. Complicated cysts will be high in signal intensity on both T1 and T2-weighted images.^{5,6} For the clinical management of small lesions which meets criteria for a cyst with benign features, serial imaging can be used.⁷ But management of larger, complicated or symptomatic lesions and parasitic cysts require surgical removal to rule out other types of adrenal neoplasms.

Adrenal cysts are usually asymptomatic but sometimes may cause abdominal, back or flank pain, fever, gastrointestinal disturbance and palpable mass, related to the size and position of the cyst. Rarely, small adrenal cysts may be associated with Cushing syndrome, virilization, or pheochromocytoma.^{2,5} One of our patients had flank discomfort and the other one had hypertension and flushing. The blood pressure levels of the latter normalized after resection of the lesion. One of the patients of Ellis et al. has a similar history of hypertension normalised after operation.²

Histopathologic findings of adrenal lymphangiomas are characterized by irregular dilated spaces lined by

flattened, simple endothelial cells. Endothelium is typically positive with vascular markers such as CD31 and CD34 immunohistochemically. Ellis et al. recommend D2-40, a transmembrane mucoprotein, which is expressed by lymphatic endothelial cells. They mention that D2-40 is a more specific marker for lymphatic lineage but D2-40 has been shown to be expressed by epithelioid-type mesothelial neoplasms so lack of cytokeratin staining should be demonstrated.² Cystic lymphangioma-like adenomatoid tumour of the adrenal gland may be included to the histopathologic differential diagnosis. These tumours are very rarely located in extragenital sites and adrenal gland. They are characterized by tubules and cleft-like spaces lined by flattened cells and lymphangioma is the main histologic mimic of adenomatoid tumour. However the lining of the spaces are immunopositive for cytokeratins and negative for endothelial markers.⁸

Conclusion

Cystic adrenal lymphangiomas are very rare benign lesions. Although previously these lesions were primarily found at autopsy, they are currently detected as incidental findings during imaging work-up for unrelated causes. They can easily imitate other adrenal neoplasms. Lymphangiomas must be kept in mind in the clinical and radiologic differential diagnosis of cystic adrenal lesions.

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