

Oncology

An adrenal cystic lymphangioma: A case report of a rare tumor

Selim Zaghibib*, Marouene Chakroun, Ahmed Saadi, Kays Chaker, Haroun Ayed, Mohamed Chebil

Department of Urology, Charles Nicolle Hospital, Tunis, Tunisia



ARTICLE INFO

Keywords:

Cystic lymphangioma
Adrenal tumor
Cystic lesion

ABSTRACT

Cystic lymphangioma is a rare benign tumor developing from lymphatic endothelial cells, and is usually described in the neck or axilla. We report a new case of an adrenal cystic lymphangioma fortuitously found in a 37-year-old patient. This case report and review of the literature bring new insights into the diagnostic difficulty and management of cystic lymphangioma of the adrenal gland.

Introduction

Cystic lymphangioma is a rare benign tumor developing from lymphatic endothelial cells. This entity is usually described in the neck or axilla, yet it may concern other sites including the abdominal cavity which accounts for 5% of all lesions.¹ Up to now, only 53 cases of cystic lymphangioma of the adrenal gland (ACL) have been reported.² We report a new case of (ACL) fortuitously found in a 37-year-old patient.

Case presentation

A 37-year-old woman, with no past medical history was referred to our department for left adrenal incidentaloma. The patient was asymptomatic, she has not experienced headache, palpitations or flank pain. Physical examination, including blood pressure, and routine laboratory examinations were normal. Abdominal computed tomography (CT) scan showed a left adrenal tumor, measuring 47.7×21.3 mm in diameter which was well-circumscribed, had a low density, with very slight enhancement, containing multiple calcifications (Fig. 1). Hormonal studies including urine metanephrine, normetanephrine and vanilmandelic acid were all normal. Also plasma cortisol concentration was normal.

The size of the lesion (> 4 cm) and the presence of calcifications were considered as risk factors for malignancy and therefore surgical removal of left adrenal gland was considered appropriate. Laparoscopic left adrenalectomy via lomboscopic approach was performed and the tumor was excised en-block with the left adrenal gland. The post-operative period was uneventful, and the patient was discharged on the second postoperative day.

The specimen received by the pathology laboratory measured

$5 \times 3.5 \times 1$ cm and the cut section revealed an adrenal mass with multiple cystic spaces with calcifications.

Microscopic examination showed a multi-cystic lesion with flat and bland endothelial cells, containing eosinophilic material, adjacent to the normal-appearing adrenal cortex (Fig. 2). Immunohistochemically, the tumor was positive for CD31 and CD 34 and the diagnosis of adrenal cystic lymphangioma was made.

The patient is now under follow up for 10 months, she is free of symptoms and no abnormality was observed.

Discussion

In 1828, cystic lymphangioma was firstly reported by Reden Backer. They are caused by a congenital obstruction or agenesis of lymphatic tissue results in lymphangiectasia due to lack of normal communication of the lymphatic system.³ Although it is commonly thought of benign tumor occurring in the neck, axilla and mediastinum observed in children, it could arise from intra-abdominal organs, especially the mesentery, in contrast to adrenal location that is very rare. Up to now, only 53 cases of occurring ACL have been reported.² Mean age at diagnosis was 39.5 (16–60) years.² Clinical examination can show a flank pain, hypertension which seems to be non-related to ACL or a palpable mass. It's remarkable that ACL were diagnosed incidentally in 15 cases.² In fact, ACLs are usually asymptomatic when they have a small size and in reported cases, mean tumor size was 8.86 (2–35) cm.² They are more common in women with no formal explanation for that female predominance. Blood tests are usually not helpful as a diagnostic tool. Although ACLs are not hormone-productive lesions, hypersecretion was reported in six cases.² Since the lesion is uncommon in this organ, imaging features lack specificity. US is a good-first exam modality and

* Corresponding author.

E-mail address: selim.zag@gmail.com (S. Zaghibib).

<https://doi.org/10.1016/j.eucr.2019.100843>

Received 1 December 2018; Received in revised form 23 January 2019; Accepted 30 January 2019

Available online 01 February 2019

2214-4420/© 2019 Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).



Fig. 1. Abdominal computed tomography (CT) scan showed a left adrenal tumor, measuring 47.7×21.3 mm in diameter which was well-circumscribed, had a low density, containing multiple calcifications.

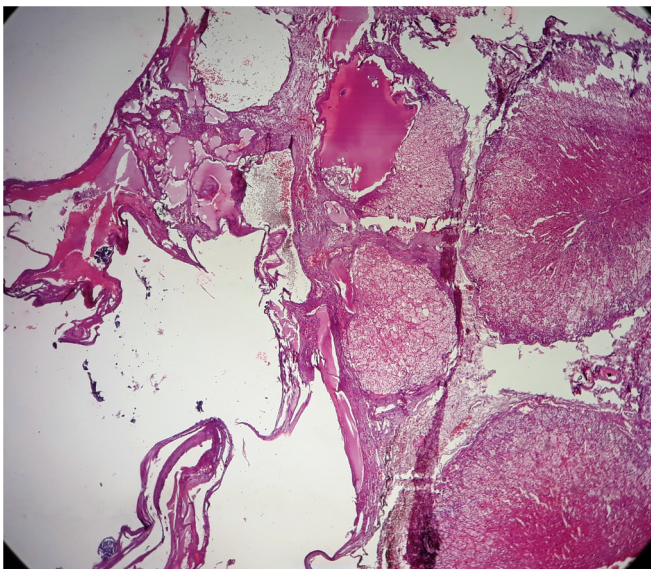


Fig. 2. Microscopic examination showed a multi-cystic lesion with flat and bland endothelial cells, containing eosinophilic material, adjacent to the normal-appearing adrenal cortex.

reveals an anechoic lesion in the suprarenal location.⁴ On CT, ACL appears hypodense with smooth borders.⁴ Although MRI is more specific than CT, T1- and T2-weighted MR images are not pathognomonic.⁴ On T1-weighted sequences adrenal lymphangioma appears as a hypointense cystic image, which borders are delineated by injection of contrast and homogeneously hyperintense on T2-weighted sequences.⁴

Clinical management of ACLs usually depends on imaging findings as lesion size is the main indication for AL removal with no consensus in the literature regarding the optimal size of the tumor in need of surgical intervention. Therefore, non-surgical treatment should be discussed in not hormone-productive small lesions especially when they are asymptomatic (16 cases including our case), with no malignancy features in imaging exams.

Histologically ACLs are distinguished by a recognizable endothelial lining, with mainly proteinaceous content of the cyst. Multiloculated cystic and endothelial lined cavities with lymphocyte aggregation are focally observed on the cyst's wall. Typical immunohistochemical characteristics are positivity for CD31, CD34 and factor VIII-related antigen and a lack of staining for cytokeratin which confirm the lymphatic rather than the mesothelial nature of these tumors.⁵

Conclusion

ACLs are rare, cystic, benign lesions. Its diagnosis can be difficult and challenging since imaging features are not specific and it is usually established after surgery by pathological report.

References

1. Ellis CL, Banerjee P, Carney E, Sharma R, Netto GJ. Adrenal lymphangioma: clinicopathologic and immunohistochemical characteristics of a rare lesion. *Hum Pathol*. 2011;42:1013–1018.
2. Michalopoulos N, Laskou S, Karayannopoulou G, Pavlidis L, Kanellos I. Adrenal gland lymphangiomas. *Indian J Surg*. 2015;77(Suppl 3):1334–1342.
3. Solomou EG, Patriarheas GV, Mpadra FA, Karamouzis MV, Dimopoulos I. Asymptomatic adult cystic lymphangioma of the spleen: case report and review of the literature. *Magn Reson Imag*. 2003;21:81–84. [https://doi.org/10.1016/S0730-725X\(02\)00624-0](https://doi.org/10.1016/S0730-725X(02)00624-0).
4. Rodrigo Gasque C, Martí-Bonmatí L, Dosdá R, Gonzalez MA. MR imaging of a case of adenomatoid tumor of the adrenal gland. *Eur Radiol*. 1999;9:552–554.
5. Ates LE, Kapran Y, Erbil Y, Barbados U, Dizdaroglu F. Cystic lymphangioma of the right adrenal gland. *Pathol Oncol Res*. 2005;11:242–244. <https://doi.org/10.1007/BF02893858>.