

Case Report

Case Report of Adrenal Lymphangioma

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Abstract

We present a very unusual surgical case of adrenal lymphangioma with a personal history of papillary thyroid carcinoma.

Case presentation

Adrenal Lymphangiomas (AL) are rare vascular-derived benign tumors. As adrenal cysts and incidentalomas, AL are tumours incidentally found during a radiological screening. To our knowledge, there are less than 60 cases of these tumours documented in the international published literature. Herein, we describe an interesting case of AL in a patient with history of Papillary Thyroid Carcinoma (PTC), followed by a review of the literature on this rare tumor. This report also describes the clinical and pathologic features of AL and illustrates these conditions. AL are usually unilateral, more frequently found on the right side, with an average size of 8.8 cm, and are more frequently found in women. Its diagnosis is frequently not established until the pathological analysis confirms its nature.

Conclusion

AL are rare vascular-derived benign tumours. These are frequently incidentally found by imaging studies. These tumours may

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be associated with other neoplasms, such as papillary thyroid carcinoma. In this report we describe a patient with the incidence of these two tumors. It is unknown whether there is a direct connection between this 2 neoplasms.

Keywords: Adrenal; Benign tumours; Cysts; Incidentalomas; Lymphangiomas

Background

Adrenal cysts are very rare lesions often detected incidentally during radiologic studies, as other incidental adrenal lesions. Frequency of these adrenal cysts on autopsy studies ranges from 0.06% to 0.18%. To our knowledge, there are less than 60 AL reported in the international published literature [1]. We hereby present a case of adrenal lymphangioma with a history of Papillary Thyroid Carcinoma (PTC).

Case Report

A 19 years-old female with a medical history of PTC Stage I (AJCC), presented to our institution for clinical follow-up. She had been previously treated at another institution with a total thyroidectomy, central and bilateral neck modified radical dissection 5 years prior to her admittance at our institution. During her initial clinical work-up, thyroglobulin was found to be elevated to 4.7 ng/mL and a cervical Ultrasound (US) showed evidence of regional recurrence. A thoraco-abdominal Computerized Tomography (CT) was done to rule out distant metastasis. On the enhanced CT scan, a 4.1×3.6×3.1 cm cystic lesion (36 HU, obtained through linear transformation of the measured attenuation coefficients) on the left adrenal gland was documented.

Due to the benign cystic appearance, lack of hyperfunctionality and size of the adrenal lesion, it was a common view to only give clinical surveillance to this patient. At the 6 months follow-up, a CT scan demonstrated a cystic lesion with increased diameters to 5×4×4 cm (Figure 1). Biochemical evaluation discarded hormonal alterations. As a result of the growing nature in the dimensions of this tumour (≥5 cm), it was decided to proceed with a laparoscopic left adrenalectomy with a lateral transperitoneal approach. Intraoperatively, a 5 cm cystic lesion from the left adrenal gland was identified and resected. The patient was discharged home on the post-operative day-3, without any complications.

Pathological analysis showed a 4.5×4 cm cystic tumor. The remaining of the adrenal gland was apparently normal under the microscope (Figures 2-4). Immunohistochemical analysis resulted positive for CD31 and D2-40 (arrows in Figure 5) and negative for CD34 (not shown). The final histopathological diagnosis was benign cystic lymphangioma of the left adrenal gland.

Discussion

AL are rare vascular-derived benign tumors [2,3]. There are only 2 cases documented with malignant transformation [1]. AL may occur at all ages with a peak incidence on the third to sixth decades of life,

with a mean age of 39.5 years [2,4]. AL are usually unilateral, more frequently found on the right side (59%), with an average size of 8.8 cm (range 2 - 13.5 cm), and are more frequently found in females (female to male ratio; 3:1) [1,3-5]. Lymphangiomas can appear in other abdominal organs such as liver, pancreas, gastrointestinal tube, gallbladder, greater omentum, mesocolon and kidneys. AL account for $\leq 1\%$ of all lymphangiomas [1,6,7]. The most common clinical characteristics of this neoplasm are summarized in table 1.

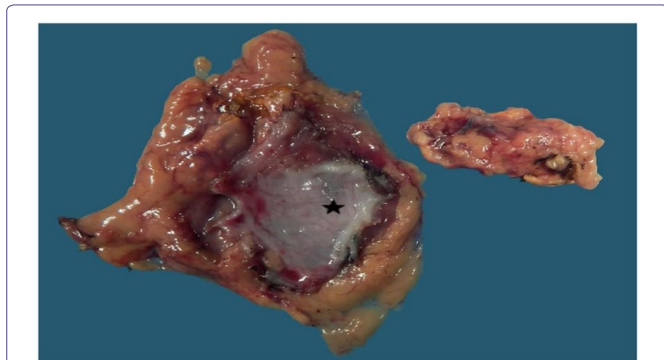


Figure 1: Abdominal CT scan: 5×4×4 cm cystic lesion (*) on the left adrenal gland, well-circumscribed, non-enhancing hypo-dense mass, with a density of 36 HU.

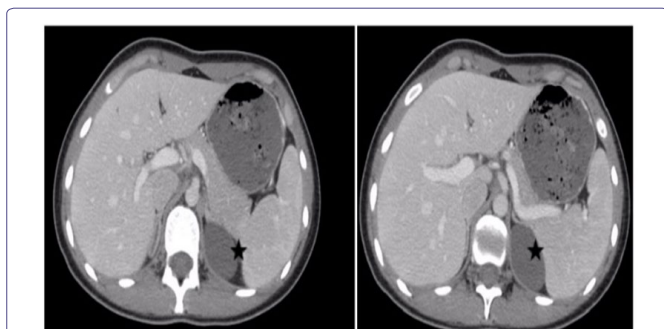


Figure 2: Pathological analysis showed that the adrenal gland had a thin, white-walled cyst (*), with a major diameter measured in 4.5 cm.

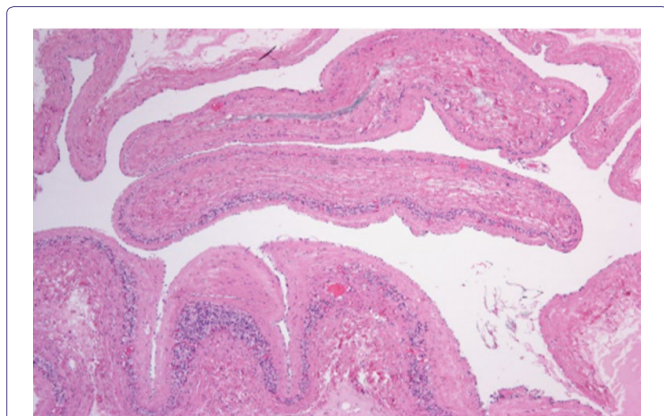


Figure 3: Microscopic view showed a complex, fibrous-walled cystic lesion with irregular locules.

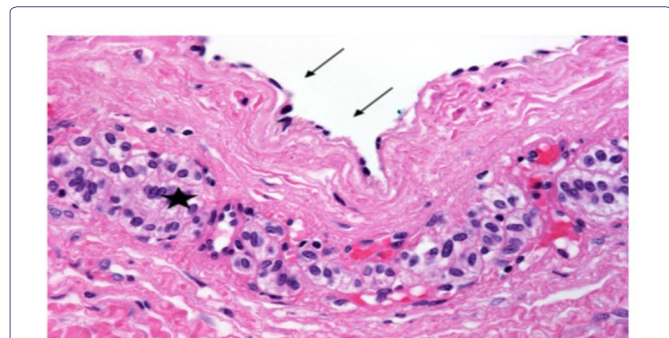


Figure 4: At higher augmentation, simple endothelial cells without atypia forming the cyst walls (arrows) with residual adrenal cortex (*) were observed.

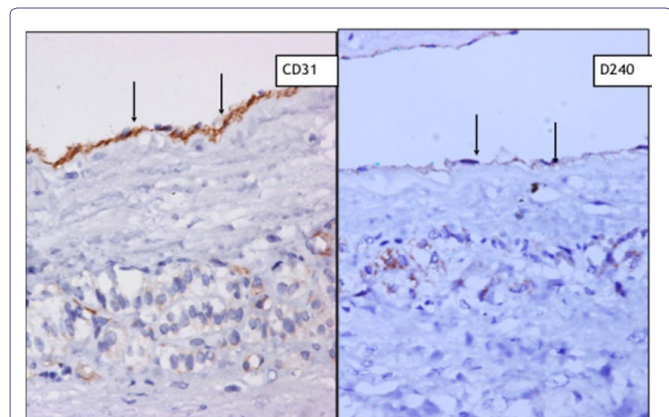


Figure 5: Immunohistochemical analysis showing positivity for CD31 and D2-40 (arrows).

Surgical Indications
Large cysts (diameter >5 cm)
High density on unenhanced CT scan (≥ 10 HU)
Delayed contrast wash-out
Significant growth (size and rate)
Hyperfunctioning cysts
Symptomatic or complicated cysts (infected, ruptured, twisted or hemorrhagic)

Table 1: Indications for surgical management of adrenal lymphangiomas [5,6,8].

AL are usually asymptomatic; when symptomatic, pain results as the foremost common symptom (44-47.9%). It can be localized in the back, flanks, and right upper quadrant or generalized [1,9]. Non-specific gastrointestinal symptoms, high blood pressure and a palpable mass can also be present. Symptoms are usually related to the mass effect.

Adrenal cysts can be classified into 4 groups: 1) Pseudocysts (39-78%), 2) Parasitic cysts (0-7%), 3) Epithelial cysts (2-9%) and 4) Endothelial cysts (20-45%). The latter can be further subdivided into angiomatous and lymphangiomatous cysts [2-5]. Rarely, adrenal cysts can be hyperfunctional tumors. There are some reports of adrenal cysts presenting as Cushing's syndrome, virilization syndrome or with elevated serum metanephrines [1,3,4,9-11].

Laboratory findings are usually nonspecific. On US cystic lymphangioma is suggested by the presence of a well-marginated, anechoic lesion, which can display some calcifications. On the CT scan, AL are identified as hypodense and non-enhancing lesions, with or without calcifications. The cystic fluid generally displays a water density or higher if hemorrhagic or a protein component are present, as seen in our patient. On Magnetic Resonance Imaging (MRI), uncomplicated adrenal cysts are low in signal intensity on T1-weighted and high on T2-weighted images [1,3,5-7].

Histopathological findings are characterized by thin-walled cystic lesions with irregular locules; filled with proteinaceous, non-viscous and eosinophilic fluid. They usually contain sheets of macrophages, lined by flattened, simple endothelial cells, which may express vascular markers, such as CD31 and D2-40. The latter is a more specific marker for lymphatic lineage [3,5]. Some authors suggest that CD34 could be positive on immunohistochemistry, while other authors mention that these lesions are negative for CD34, calretinin and cytokeratin [1,3,5,7]. In all the reported cases the diagnosis was made after the surgical removal by histological and immunohistochemical examinations [1].

The medical management of adrenal cysts is based on biochemical and clinical findings. Small asymptomatic non-functioning cysts can be treated conservatively with serial imaging [3]. The indications for the surgical intervention of adrenal lymphangiomas are shown in table 1 [5,8,9]. In our case, it was decided to proceed with the surgical management due to the growing nature of the mass and the feasibility of a safe laparoscopic approach. However, an expectant approach could have alternatively been chosen based on the cystic characteristics of the mass. So far, there are no reports of recurrence on surgically treated patients with these tumours [7].

Conclusion

AL are rare, cystic, vascular, benign and generally asymptomatic lesions. Their diagnosis is commonly established after surgery by the pathological report. Laparoscopic adrenalectomy can be safely performed and it should be offered to those AL with clinical symptoms, complications, a diameter ≥ 5 cm or with a significant growth by imaging. In this report we describe a patient with both a papillary thyroid carcinoma and an AL. It is unknown whether there is a direct connection between this 2 neoplasms.

Competing Interest

The authors declare no conflicts of interest.

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