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Endothelial cyst of the adrenal gland: A rare case report

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Abstract

Adrenal gland cysts are rare and uncommon manifestations. Mostly asymptomatic, discovered incidentally during radiological studies or at autopsy, or without characteristic symptoms. The spectrum of these entities may include benign cysts or malignant cystic neoplasms. They are classified into four types: pseudocysts, endothelial cysts, epithelial cysts, and parasitic cysts. Though pseudocysts are reported to be the most frequently clinically recognized adrenal cysts in surgical series, endothelial cysts are more frequent in autopsy series. Even with advanced imaging modalities, it is still difficult to differentiate a benign adrenal neoplasm from a malignant one. As a result, getting a definitive diagnosis and starting treatment is challenging. In both symptomatic and asymptomatic cases with a large diameter or increasing sizes during follow-up, or with any abnormality of adrenal hormones, surgery is the treatment of choice in symptomatic. Herein, we present a 47-year-old female with a nonfunctional left adrenal endothelial cyst, who was incidentally found during a computerized tomography scan. The patient presented with left-sided nephritic colic due to renal lithiasis. The objective of this paper is to recall the clinical characteristics and to specify the diagnostic contribution of imaging as well as the therapeutic modalities of this entity.

Keywords

Adrenal gland, adrenal cyst, calcification, CT-scan, adrenalectomy

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Introduction

Adrenal gland cysts are rare, with an incidence of approximately 0.06% in the general population.[1] Mostly asymptomatic, discovered incidentally during radiological studies or at autopsy, or without characteristic symptoms. They can range from benign cysts to malignant cystic neoplasms. They are classified as pseudocysts, endothelial cysts, epithelial cysts, or parasitic cysts. Though pseudocysts are reported to be the most frequently clinically recognized adrenal cysts in surgical series, endothelial cysts are more frequent in autopsy series. Even with advanced imaging modalities, it is still difficult to differentiate a benign adrenal neoplasm from a malignant one. As a result, getting a definitive diagnosis and starting treatment is challenging.[2] Management algorithms for adrenal cysts also vary and are controversial because of the overall rarity of such lesions,[3] but Surgery remains the treatment of choice in symptomatic cases as well as asymptomatic cases with a large diameter or increasing dimensions during follow-up or with any abnormality of adrenal hormones.[4] Herein, we present a 47-year-old female with a nonfunctional left adrenal endothelial cyst, who was incidentally found during a computerized tomography (CT) scan. The patient presented with left-sided nephritic colic due to renal lithiasis. This paper aims to recall the clinical characteristics and to specify the diagnostic contribution of imaging as well as the therapeutic modalities of this entity.

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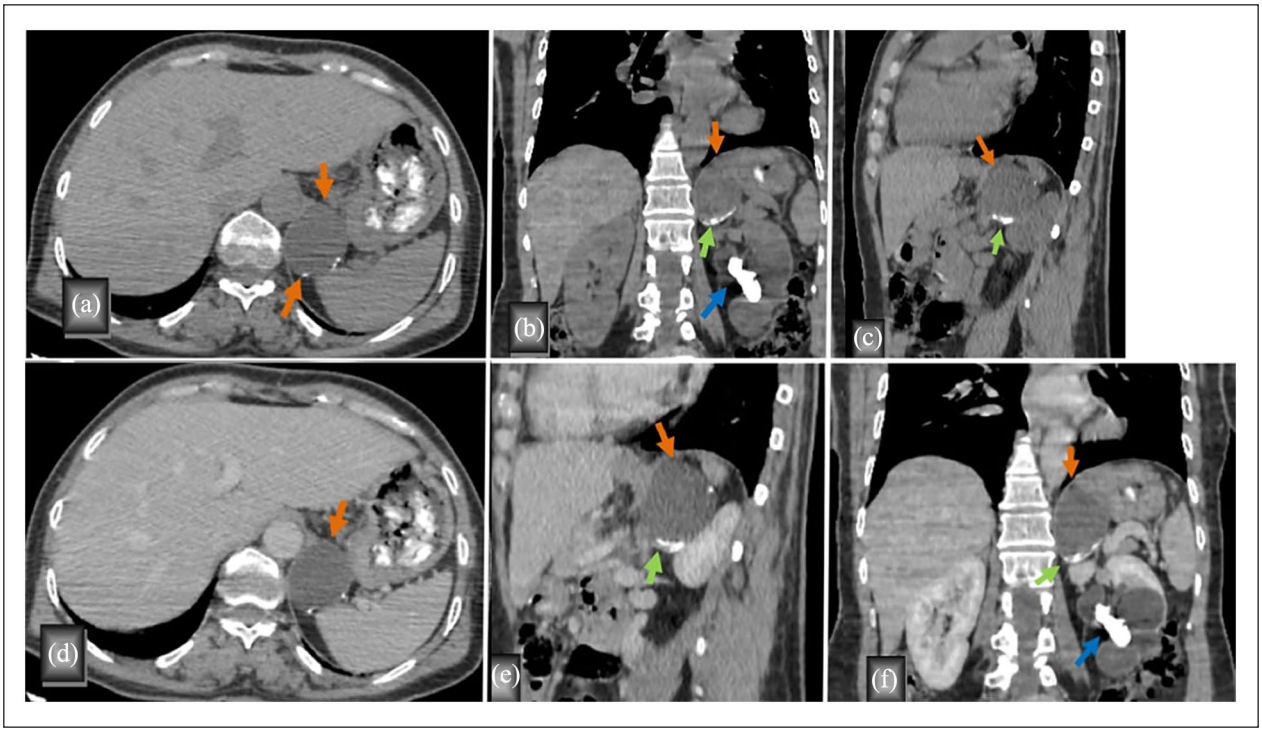

Figure 1. Non-contrast-enhanced abdominal computerized tomography scan images (a: axial, b: coronal, and c: sagittal), and enhanced images (d: axial, Figure e: sagittal, and Figure f: coronal), revealing a large well-limited cystic, homogeneous mass of the left adrenal gland (orange arrow), with regular contours, containing some linear parietal calcifications (green arrow), not septated, not enhanced after injection of contrast agent, without any rupture or intracystic hemorrhage. The mass measured 4.7 × 4.8 × 5.6 cm in cranio-caudal, anteroposterior, and transverse dimensions and was abutting various parts of the gastrointestinal tract and the kidney.
Note. Left pyelic coral calculus responsible for pyelocalyceal dilation (blue arrow).

Case report

A 47-year-old female presented with a complaint of a left nephritic colic, for more than 2 days. The clinical examination found a patient in good general condition, apyretic, with no palpable abdominal or lumbar mass, and an unremarkable cardiovascular examination. Her past medical and surgical history was otherwise uneventful, and she had no history suggestive of a functional adrenal mass. A CT scan of the abdomen was performed (Figure 1), which revealed a left pyelic coral calculus responsible for pelvicalyceal dilation associated with a large well-defined round to oval, hypodense lesion with a hyperdense non-enhancing thin-walled cystic lesion in the suprarenal region on the left side, located between the stomach and spleen, leading to significant mass effect to adjacent viscera, without any rupture or intracystic hemorrhage. There was subtle calcification in the inferior aspect; however, no enhancing solid component was identified. The mass measured 4.7 × 4.8 × 5.6 cm in craniocaudal, anteroposterior, and transverse dimensions and was abutting various parts of the gastrointestinal tract and the kidney. In light of these findings, the endocrinology team was consulted. They demanded a serum morning cortisol (6.50 mcg/dl, reference range is 5–23 mcg/dl), a 24-h urinary metanephrine (85.51 mcg, normal level <350 mcg/24 h), and nor metanephrine (177.24 mcg, normal level <600 mcg/24 h) levels which were all within normal limits.

Due to the size and position of the cyst, and to avoid a rupture of the cyst during laparoscopy, a left subcostal laparotomy was performed. During surgery, the adrenal origin of the cyst was confirmed, and an "en bloc" left adrenalectomy was performed without rupture of the cyst. No drain was left in place. The resected adrenal cyst was sent for histopathological examination.

On gross examination, the specimen measured 7 × 5.5 × 3 cm and its outer surface was smooth and congested. On serial sectioning, a citrine-yellow liquid with necrotic and calcified material was produced. The cut surface showed an uniloculated cyst measuring 7 × 5 × 3 cm. Adrenal tissue could also be identified in places. Microscopic examination revealed an endothelial cyst with a fibrocollagenous wall lined with flattened cells, with remnants of tissue adrenocortical tissue, which was normal without any evidence of malignancy (Figure 2). The surgery was performed without any intraoperative incidents and with simple postoperative follow-up. One week after the surgery, the endocrinology team was consulted about hydrocortisone replacement therapy. They ordered a serum morning cortisol which was normal at 11 mcg/dl. They concluded that the

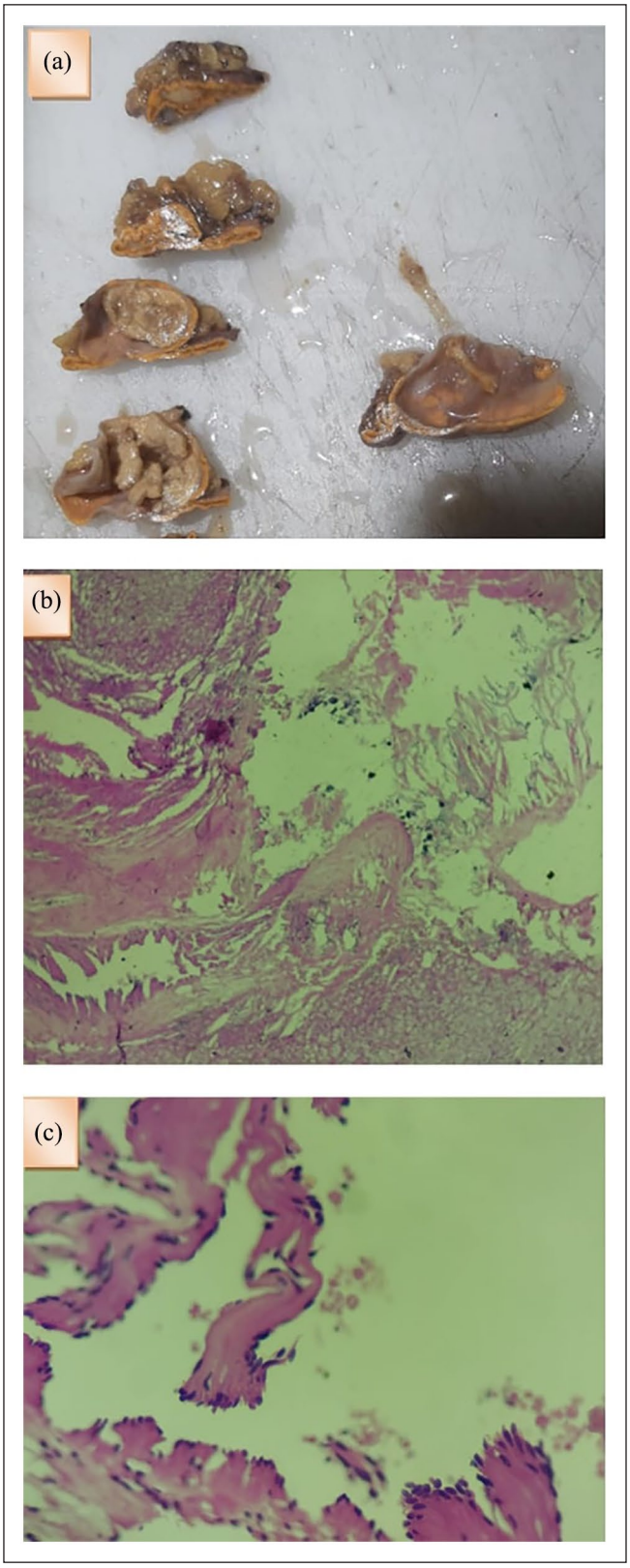

Figure 2. Adrenal endothelial cyst with a delicate lining of endothelial cells (Cut section of the specimen (a), H & E, low (b), and high (c) power).

contralateral adrenal gland functioned correctly and that replacement therapy was not needed. The patient was then discharged with a 1-month follow-up serum morning cortisol which was again normal (12.25 mcg/dl). The urology team then performed a Percutaneous Nephrolithotomy for the left pyelic coral calculus. The intervention was successful with a complete resolution of clinical symptoms. The patient is scheduled for a CT Scan in her 6-month follow-up consult.

Discussion

Adrenal gland cysts were first described by Greiselius in 1670.[5] Although adrenal cysts occur at all ages, there is a peak in incidence between the third to sixth decade of life,[6,7] with female predilection, the male-to-female ratio is 1:3.[6] The size of cysts ranges from several millimeters to more than 20 cm in diameter.[6] They are usually unilateral, but in 8%–15% they are bilateral, with equal involvement of both adrenal glands.[6,8,9] Four main types are distinguished based on histological examination and incidence[1]: Endothelial cysts (45%) of lymphangiomatous or angiomatous origin, pseudocysts, often highly vascularized and hemorrhagic (39%), epithelial cysts (9%), and cysts of parasitic origin, often hydatid (7%). Endothelial cysts are postulated to be formed by dilated and thrombosed vessels with organization.[10] The anatomopathological examination of the surgical specimen of our case concluded with an endothelial cyst without signs of malignancy. Endothelial cysts, also known as simple cysts, are the most common among adrenal cysts in autopsy series with an incidence of 45% but account for only 2%–24% of clinically symptomatic lesions.[11]

Clinically, because most adrenal cysts are asymptomatic, they are usually found as incidental findings on imaging studies or incidentally during surgery performed for other abdominal pathologies. Symptoms, usually related to the size, and position of the cyst, may include pain, gastrointestinal disturbance, or palpable mass. Hypertension is another rarely observed symptom in 9% of cases due to compression of the adrenal artery or renal medulla.[9] More rarely, they may be the cause of an acute abdominal pain syndrome due to an intracystic hemorrhage or a state of shock due to retroperitoneal hemorrhage.[1]

Radiology

Although radiological studies can be useful for the pre-surgical diagnosis of cystic lesions of the adrenal gland,[2] they are usually inadequate for the definitive histologic type of a cystic adrenal lesion, or to distinguish the benign from malignant nature of the disease.[4,7] Therefore, in any case of suspicion further investigations, biopsies, or surgery are usually performed to rule out malignancy. Endothelial cyst of the adrenal gland is characterized on the ultrasound (US), as cysts similar

to those seen elsewhere in the body. They are well-defined and anechoic homogenous with posterior acoustic enhancement, with thin and regular walls in the US. Sometimes they do not appear homogenous and may have septations or foci of internal echogenities, which are usually caused by hemorrhages.[7,8] Internal septa are better demonstrated by the US than by CT scan. The CT scan is usually the primary imaging modality for both the detection and differentiation of adrenal masses. Specially dedicated thin-sliced images with coronal reconstruction depict the anatomical relationships.[6] Adrenal cysts are of fluid density with well-defined margins and thin walls on CT scans. There is no enhancement after intravenous injection with contrast medium. Calcification, which is often peripheral and curvilinear, is seen in 15% of adrenal cysts. Higher density within the cyst may occur due to hemorrhage.[7] In our case, the CT scan confirmed the cystic nature of the adrenal mass, with peripheral parietal calcification, and no enhancement after contrast administration. A lack of enhancement helps to differentiate adrenal cysts from other adrenal lesions such as adenomas. A complex cyst may be difficult to differentiate from metastasis or other necrotic tumors or abscesses. Cysts may be difficult to attribute to the adrenal gland when they are very large. The multiplanar capability of magnetic resonance imaging (MRI) and US helps localize a large mass to the adrenal.[8]

Histopathology

On gross examination, endothelial cysts are well circumscribed and surrounded by a capsule, with sizes ranging from 1.4 cm to 33 cm. Microscopically, endothelial cysts comprise of fibrous wall with patchy lining by flattened cells resembling normal endothelium.[6] These cells are positive for endothelial cell markers that is, podoplanin, FLI1, CD31, and factor VIII, confirming their endothelial nature. Clinically, differential diagnoses of vascular adrenal cysts include cystically degenerated adrenal neoplasms, such as pheochromocytoma, adrenal cortical adenoma, adrenal cortical carcinoma, or rarely metastatic carcinoma. Detailed grossing including a close examination of cystic contents with a meticulous sampling of the wall can help reach a conclusive diagnosis.[6,12]

Management

True adrenal cysts are usually non-functional tumors in terms of hormone secretion.[13,14] Nonetheless, necrosis or intratumor hemorrhage can cause primary and secondary adrenal tumors to change into cysts. Therefore, it is important to always consider the potential hormonal functionality and malignancy of adrenal cystic lesions. Cysts measuring more than 5 cm are considered at a higher risk of malignancy and are usually presented as pseudocysts.[15] Our patient's cyst had a size of 5.8 cm in its transverse dimension and benefited from hormonal screening. The first and most important step in evaluating any adrenal tumor, even cystic ones, is to rule out pheochromocytoma.[16] The current guidelines state that the initial testing for pheochromocytoma includes the biochemical analyses of catecholamine metabolites: urine-fractionated metanephrines or plasma-free metanephrines. Normal biochemical testing, however, does not completely rule out the potential of pheochromocytomas, as up to 30% of individuals could have a non-secretory or a clinically silent pheochromocytoma.[13]

The 1 mg dexamethasone suppression test is the second required test for assessing all adrenal tumors. It is conducted to rule out autonomous cortisol secretion. According to the most recent guidelines for managing adrenal incidentalomas, all patients with incidentally discovered adrenal masses should have this evaluation regardless of the clinical signs of hypercorticism.[17] Testing for excess androgens and mineralocorticoids in patients with adrenal tumors is not obligatory if there is no clinical suspicion.[17] Primary hyperaldosteronism in cases with unilateral adrenal masses is usually due to small adrenal tumors (<2 cm). To our knowledge, no cases of benign cystic aldosterone-producing adenomas have been reported.[18] To date, in the largest cohort study of patients with benign adrenal cysts, primary hyperaldosteronism was suspected in 4% of patients, but further testing has not verified the suspicion.[14] Thus, aldosterone-secreting adrenocortical carcinoma (ACC) should always be suspected when hypertension and hypokalemia coexist with a large cystic adrenal tumor.[18] Similarly, in the case of a large cystic adrenal tumor, hirsutism, and/or virilization may be suggestive of the presence of a pure androgen-secreting adrenocortical tumor, of which 50% are typically ACCs.[19] The latter are rare malignant masses. Their typical appearance on imaging is a large, heterogeneous solid tumor with irregular margins infiltrating adjacent anatomical structures.[20]

Optimum management of adrenal cysts remains controversial, due to their low incidence. Surgical management, open or minimally invasive, depends on a surgeon's preference, tumor characteristics, and expertise. Functional cysts, malignant or possibly malignant cysts, symptomatic cysts of any size, asymptomatic cysts larger than 5 cm, and patients with unclear follow-up are typically candidates for surgery.[1,4,21] Conservative management is apt in those with uncomplicated/asymptomatic cysts <5 cm. In our case, the mass was larger than 5 cm; thus, the surgical team proposed surgical management which was approved by our patient. Though the postoperative period of such patients is uneventful, and they recover well, a minimum of 18 months of follow-up, with repeat CT scans every 6 months is indicated. Aspiration of cysts can be considered as an alternative to surgery in the case of surgically unfit patients.[4] Marsupialization or decortication have also been tried as alternatives to surgery for large cysts especially those cysts that are adherent to multiple organs where excision may be difficult. Sclerotherapy using absolute alcohol has also been described but it is associated with a high recurrence of 30%–50%.[4,22]

Conclusions

Adrenal cystic lesions are uncommon and can range from benign cysts to cystic primary adrenal malignancy. Although it is challenging to differentiate an adrenal cyst from other adrenal neoplasms, imaging studies, including CT scans and MRI, are essential for defining an adrenal cystic lesion and differentiating it from a cystic lesion of adjacent organs. All cystic adrenal lesions require a histological examination since benign cysts can mask underlying malignancy. In both symptomatic and asymptomatic cases with a large diameter or increasing sizes during follow-up, or with any aberration of adrenal hormones, surgery is the optimal method of treatment.

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Author contributions

All authors contributed to this work. All authors have read and approved the final version of the manuscript.

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Informed consent

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