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CASE REPORT

Adrenal endothelial cyst associated with adrenocortical adenoma: a case report and literature review

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Abstract

We present a case of a middle-aged male Chinese patient who was asymptomatic with a large (6 × 7 cm) right adrenal mass was found in this patient upon routine health examination. He underwent laparoscopic right adrenalectomy after comprehensive evaluation, and the mass was finally diagnosed as right adrenal endothelial (vasculature) cyst associated with adrenocortical adenoma according to pathological and immunohistochemical studies. The puzzling image resemblance of a variation of adrenal cyst to carcinoma necessitated histological examination for confirmative diagnosis. The development of endothelial cyst is extremely rare, and its association with other adrenal neoplasms is even rarer. Herein, we report a new case of adrenal endothelial cyst associated with adrenocortical adenoma, which was almost indistinguishable from adrenocortical carcinoma, and hope that it would be helpful in the diagnosis of other similar cases.

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Key words: adrenal; endothelial cyst; adenoma; urology

A 53-year-old man was hospitalized because of a large right adrenal mass that was incidentally found on plain and contrast-enhanced abdominal computed tomography (CT) scan upon routine health examination. The patient has type 2 diabetes for > 2 years, and blood glucose level was well controlled with regular use of glimepiride (4 mg once daily) and acarbose (50 mg thrice daily). He had no significant medical or family history. He did not show any Cushingoid features, such as moon face and skin atrophy, on physical examination. Body mass index was 26.1 kg/m^2 (height, 1.65 m; weight, 71 kg), and the pattern of fat deposition was generalized rather than centripetal. The preoperative blood pressure ranged from 114/78 mmHg to 133/89 mmHg, and the heart rate was from 73 beats/min to 88 beats/min. The laboratory examination results, including biochemical and hematological investigations, urinalysis, and coagulation, were within normal limits. Basal plasma levels of prostatic antigen Total Prostate Specific Antigen (TPSA) and Free Prostate Specific Antigen (FPSA), cortisol, aldosterone, urine 17-ketosteroid, and 3-methoxy 4-hydroxymandelic acid were within normal limits (Table 1). Abdominal CT scan (Fig. 1) demonstrated a huge (6 × 7 cm) circular low-density right adrenal mass. A multi-room separation can be observed inside the mass, and plaque-like high-density calcification can be observed on the wall of the capsule. The CT value is approximately 17 HU. There was no significant enhancement on contrast-enhanced CT. After comprehensive condition assessment, case discussions, and obtaining informed consent from the patient, the possibility that the tumor may be a nonfunctioning adrenocortical carcinoma (AC) was considered. Laparoscopic right adrenalectomy was performed. The specimen was sent for histopathological and immunohistochemical examination. The cystic mass (7.0 × 6.0 × 2.5 cm) has clear content, and the pale-yellow mass (5.0 × 1.0 × 1.0 cm) was noted outside the capsule wall, whose slice surface was yellowish and soft (Fig. 2). Microscopically, the cystic mass was composed of fibrous sac wall tissues with local calcification. Another mass with fibrous capsule outside the cystic wall

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Table 1 Summary of the preoperative laboratory data

Items	Unit names	Measured value	Range
TPSA	ng/mL	0.54	(0–4)
FPSA	ng/mL	0.2	(≤ 4)
Cortisol (4:00 pm)	μg/dL	4.9	(1.5–12)
Cortisol (12:00 am)	μg/dL	1.74	(1.5–8)
Cortisol (8:00 am)	μg/dL	8.19	(4.3–22.4)
Aldosterone (standing position)	ng/L	164.8	(65–295)
Aldosterone (supine position)	ng/L	128.95	(59–173)
17-OHCS	μmol/24h	9.1	(8.3–27.7)
VMA	μmol/25h	16	(0–68.6)
CEA	μg/L	1.9	(0–5)
AFP	μg/L	3.5	(< 9)
CA-125	IU/mL	4.6	(0–68)
CA-199	U/mL	16.87	(0–37)

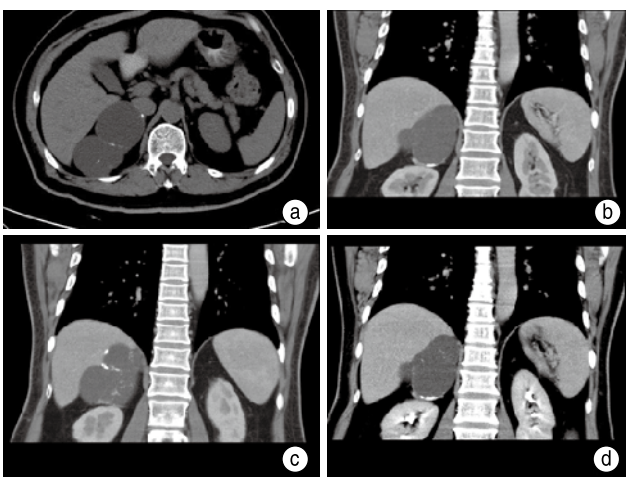

Fig. 1 Transversal CT scanning. (a) 6 × 7 cm circular right adrenal mass. Coronal CT scanning; (b) Low-density mass, CT value 17 HU; (c) Multi-room separation inside the mass; (d) Plaque-like high-density calcification on cyst wall

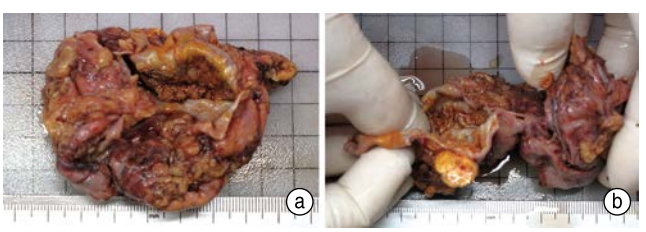

Fig. 2 (a) Cystic mass (7.0 × 6.0 × 2.5cm) with clear content; (b) Pale-yellow mass (5.0 × 1.0 × 1.0cm) outside the capsule wall, with yellowish and soft slice surface

composed of bright and dark cells, which are arranged in acinar and flaky shapes, can be found, and a large and deformed nucleus was present in the foci. The result of immunohistochemistry shows that the cells stained positive for CD34 (angiomatous marker/+), D2-40 (lymph angiomatous marker/+), desmin(+), and SMA(+) (Fig. 3).

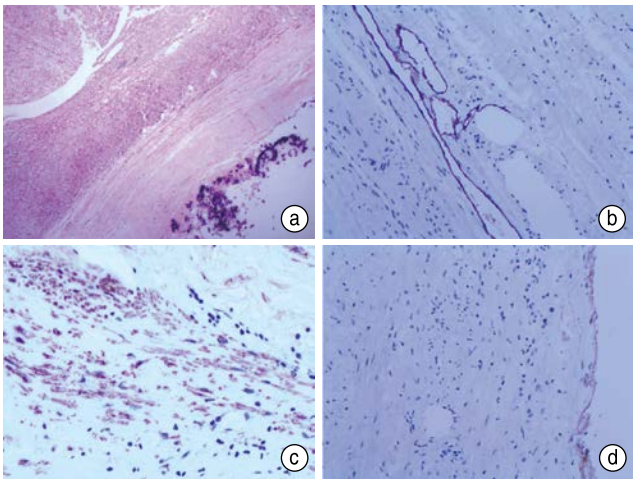

Fig. 3 Microscopically (HE staining × 40), the junction of fibrous sac wall tissue and adrenocortical adenoma cells (a). The result of immunohistochemistry shows that the cells stained positive for D2-40 (b), SMA (c), and CD34 (d)

Based on the histological and immunohistochemical results, right adrenal endothelial (vasculature) cyst associated with adrenocortical adenoma was the final pathologic diagnosis. The patient had an uneventful hospital course and was discharged on the 13th postoperative day.

Discussion

Incidental adrenal masses are common and develop in approximately 3%–7% of adults. They are more frequently detected because of increased utilization and improved spatial resolution of CT and magnetic resonance imaging (MRI). The pathological types of adrenal tumors include adenomas, myelolipomas, pheochromocytomas, ACs, adrenal lymphoma, and cysts. The most frequent type is a benign, non-hyperfunctioning adenoma [1].

An adenoma is usually incidentally found on imaging and exhibits no hormonal activity. When hormonally active, it may produce cortisol or aldosterone. Adenomas can be accompanied by the following clinical syndromes: hypercortisolism (Cushing's syndrome) and hyperaldosteronism [2]. Generally, adenomas can be divided into two groups: lipid-rich and lipid-poor adenomas. This determines the appearance of adenomas on CT and MRI [3].

Cysts are a rare pathological type of adrenal glands. The number of patients with adrenal cystic lesions determined by autopsy is approximately 14 per 20,000, which constitutes 0.074% of the general population [4]. Since most adrenal cysts are asymptomatic, they are usually incidental findings on imaging studies or discovered incidentally during surgery performed for

other abdominal pathologies. Their diagnostic rate has increased in the last two decades due to the widespread use of imaging studies with improved techniques [5]. The sensitivity of a CT for an abdominal hydatid cyst is 97%. To identify a hydatid cyst, CT or MRI can reveal a cystic lesion and the presence of daughter cysts [6]. The radiological features of cystic lesions are diverse because of the heterogeneous texture within the cysts. Classifications have been proposed, and that by Barron and Emanuel in 1961 is now widely accepted. They categorized adrenal cystic lesions into pseudocysts, epithelial cysts, endothelial cysts, and parasitic cysts [7].

The development of endothelial cyst is extremely rare, and its association with other adrenal neoplasms is even rarer. The review by Erickson et al. included only one case of endothelial cyst associated with pheochromocytoma [8]. In 2009, Nigawara *et al*. reported the first case of adrenal endothelial cyst associated with adrenocortical adenoma [9]. Herein, we report a second similar case of adrenal endothelial cyst associated with adrenocortical adenoma, which was almost indistinguishable from AC.

The main challenge of managing adrenal mass is to correctly distinguish the rare unexpected malignant lesion or hyperfunctioning adenoma from a benign, clinically insignificant disease. Adrenalectomy is required in hormonally active tumors responsible for syndromes and symptoms of uncontrolled release of hormones. The Adrenal Subcommittee of the American College of Radiology Incidental Findings Committee has presented the algorithm for evaluation of an incidentally detected adrenal mass. The algorithm was applied to patients who are aged > 18 years, asymptomatic, and referred to imaging for a reason that is unrelated to potential adrenal pathology. Moreover, it recommended that patients with adrenal mass > 4 cm in size without cancer history may consider resection [1]. Smaller tumors, growing over the period of observation, or those that do not fulfill the criteria of benign adenoma in imaging studies should also be excised [4].

Therefore, we presented a case of adrenal endothelial cyst associated with adrenocortical adenoma, which could not be clearly discriminated from AC in preoperative imaging studies. Suspicious large adrenal mass (> 4 cm) should always be closely characterized and considered for resection because it may compress surrounding organs. Moreover, there is a potential risk of malignancy that requires detailed histological exploration. When we do not attempt to distinguish clinically significant from insignificant disease, we are at risk for overdiagnosis or even misdiagnosis.

Conflicts of interest

The authors indicated no potential conflicts of interest.

References

1. Mayo-Smith WW, Song JH, Boland GL, *et al*. Management of incidental adrenal masses: a white paper of the ACR incidental findings committee. J Am Coll Radiol, 2017, 14: 1038–1044.
2. Ctvrtlik F, Koranda P, Tichy T. Adrenal disease: a clinical update and overview of imaging. A review. Biomed Pap Med Fac Univ Palacky Olomouc Czech Repub, 2014, 158: 23–34.
3. Korobkin M, Giordano TJ, Brodeur FJ, *et al*. Adrenal adenomas: relationship between histologic lipid and CT and MR findings. Radiology, 1996, 200: 743–747.
4. Major P, Pędziwiatr M, Matłok M, *et al*. Cystic adrenal lesions - analysis of indications and results of treatment. Pol Przegl Chir, 2012, 84: 184–189.
5. Chien HP, Chang YS, Hsu PS, *et al*. Adrenal cystic lesions: a clinicopathological analysis of 25 cases with proposed histogenesis and review of the literature. Endocr Pathol, 2008, 19: 274–281.
6. Akçay MN, Akçay G, Balik AA, *et al*. Hydatid cysts of the adrenal gland: review of nine patients. World J Surg, 2004, 28: 97–99.
7. Barron SH, Emanuel B, Adrenal cyst. A case report and a review ok the pediatric literature. J Pediatr, 1961, 59: 592–599.
8. Erickson LA, Lloyd RV, Hartman R, *et al*. Cystic adrenal neoplasms. Cancer, 2004, 101: 1537–1544.
9. Nigawara T, Sakihara S, Kageyama K, *et al*. Endothelial cyst of the adrenal gland associated with adrenocortical adenoma: preoperative images simulate carcinoma. Inter Med, 2009, 48: 235–240.

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