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Case Report

Adrenal cyst – A diagnostic quandary

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ABSTRACT

Adrenal cysts are rare benign entities that commonly masquerade as more serious illnesses. They are usually asymptomatic and non-functional. Here, we present a case of a large left adrenal cyst of size 10.8 × 10.2 × 7.8 cm in a 25-year-old male, who initially presented with fever and abdominal pain, with clinical suspicion of an abdominal malignant neoplasm. Laboratory workup was unremarkable, while the histopathological diagnosis was that of a benign true adrenal cyst – epithelial type. This case report highlights the rarity of cysts larger than 4 cm, radiological and pathological features for appropriate diagnosis, as well as subclassification of the cyst, which is essential to institute optimal treatment.

Keywords: Adrenal cyst, Epithelial cyst, Benign, Adrenalectomy

INTRODUCTION

Adrenal cysts are rare masses, with an incidence of <1%, that arise from the adrenal gland. Most of these are non-functional and asymptomatic, usually measuring <10 cm and, on average, are about 4 cm. Larger cysts may present with abdominal distension. They may occur in conjunction with other adrenal neoplasms. Here, we report a rare case of a large true adrenal cyst – epithelial type, which is a rare retroperitoneal cyst that seldom reaches sizes sufficient to manifest clinically such as in our case.

CASE REPORT

A 25-year-old male presented to the urology department of our hospital with fever, abdominal fullness, and pain for 1 month. Laboratory investigations showed normal values of blood urea nitrogen, creatinine, sodium, potassium, chloride, bicarbonate, calcium, and albumin. On further evaluation, an abdomen mass was detected on USG. Following this, he was further subjected to a CT urogram, which revealed a left-sided hypoechoic cystic adrenal lesion of 10.8 × 10.2 × 7.8 cm [Figure 1]. A hormonal workup was also performed which was unremarkable. An adrenal cyst was suspected and the lesion was resected and the patient was hemodynamically stable after the procedure.

The specimen was sent for histopathological examination and grossly showed a single grey-white flap-like soft-tissue fragment measuring 8 × 2 cm. The external surface was grey-white and unremarkable, while the inner surface was grey-white with focal grey-yellow areas. No solid or papillary areas were identified. Random bits from the soft-tissue fragment and cyst wall were partially embedded.

Microscopic examination revealed sections of the adrenal gland with a clearly demarcated cyst wall [Figure 2], lined by flattened epithelium and congested blood vessels in all the sections examined [Figures 3-5].

A final histopathological diagnosis was a benign simple left adrenal cyst – epithelial type.

DISCUSSION

Adrenal cysts are uncommon lesions, with an incidence of 0.06–0.18%, and account for about 4% of adrenal masses.[1] They may occur at any age, although they are more frequent in young adults with a slight female predilection. The size of the cysts may vastly vary from a few millimeters to over 4 cm, with very few cysts exceeding 10 cm. In addition, they are commonly solitary, and affect both the right and left adrenal glands with equal incidence. They may either be uniloculated or multiloculated.[2,3]

While the routine clinical presentation largely depends on the location and largest dimension of these benign cysts and is usually asymptomatic [Table 1], other features such as dyspnea, abdominal pain, gastrointestinal disorders, and occasionally, a palpable mass may also be seen in large cysts. Rarely, intracystic hemorrhage and infection, as well as hypertension have been also reported.[2] Our patient presented

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only with vague abdominal pain despite a cyst of size 10 cm in the greatest dimension, attributing to a rare presentation. In our case, while the CT scan was indicative of an adrenal cyst [Figure 1], neoplastic lesions could not be ruled out clinically due to the size and increased attenuation coefficient of the lesion, warranting a histopathological examination in this case.

Adrenal cysts were originally classified into four major histopathological types: Epithelial cysts, parasitic cysts, (specifically echinococcal type), pseudocysts, and endothelial cysts which were the most common subtype.[1,4-6] Among these, true epithelial adrenal cysts are even rarer, accounting for only 9% of all adrenal cysts. This has further been adopted in the latest WHO classification, which has introduced a new chapter on adrenal cysts. It also takes into account, the potential of simulating primary cystic adrenal neoplasms or adrenal cortical carcinomas in the setting of an adrenal pseudocyst, which would invariably lead to a misdiagnosis.[7]

The presence of a cyst lining is assessed first, if absent, a diagnosis of an adrenal pseudocyst is favored, which generally contains a fibrinohemorrhagic fluid and is demarcated by thickened fibrous tissue. If a lining is present, it is to be further assessed if the cyst contains a monolayer of endothelial or mesothelial cells, or flattened epithelial cells. The flat endothelial cells are associated with a lymphatic lining and occasional intraluminal papillary projections may be observed. The mesothelial cells usually lack atypia but may be either flat or cuboidal.

Parasitic cysts frequently grow to very large sizes and contain clear fluid in uniloculated or multiloculated cystic spaces. They are usually enclosed by a fibrotic and calcified wall with parasitic membranes demonstrating PAS positivity.[8-10]

The described microscopic features generally do not pose diagnostic challenges with these cystic lesions in the absence of solid areas or cell atypia. Extensive sampling of the cyst wall is mandatory to rule out an underlying neoplasm, as some adrenal cortical neoplasms or pheochromocytomas with cystic degeneration with hemorrhage may mimic benign adrenal cysts.[11]

True epithelial cysts, such as in our case, have a lining composed of flattened epithelial cells, which may be confirmed by IHC staining for Cytokeratin (AE1/AE3). Pathological examination of the resected tumor in our case was suggestive of a diagnosis of an epithelial cyst, which is characterized by a cystic cavity with a surrounding fibrous wall, and multilocular spaces lined by epithelial cells. Morphology is the key to diagnosis and IHC workup may be suggested in partially lined cysts or equivocal cases, and hence, was not warranted in our case.[12]

Endothelial cysts are described radiologically as unilateral, thin-walled (usually <3.5 mm), variably sized with smooth borders, multilocular structures, and occasional septal calcifications.

Epithelial cysts while they usually have a low attenuation coefficient of <20 Hounsfield units, the cystic contents are more prone to hemorrhage than any other subtype and may cause an increase in the attenuation coefficient.[2]

Endothelial cysts, usually referred to as simple cysts, are frequently multiloculated and are thin-walled measuring <2 cm in size. They are lined by a flat endothelial lining and may contain a yellow-tinged serous fluid that may be admixed with hemorrhage. They can be further subclassified

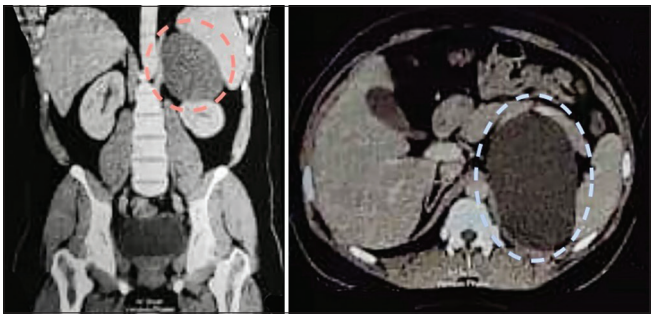

Figure 1: CECT Abdomen shows a left sided hypoechoic cystic adrenal lesion measuring 10.8 × 10.2 × 7.8 cm in AP (left) and transverse (right) views, denoted by red and blue dotted lines, respectively.

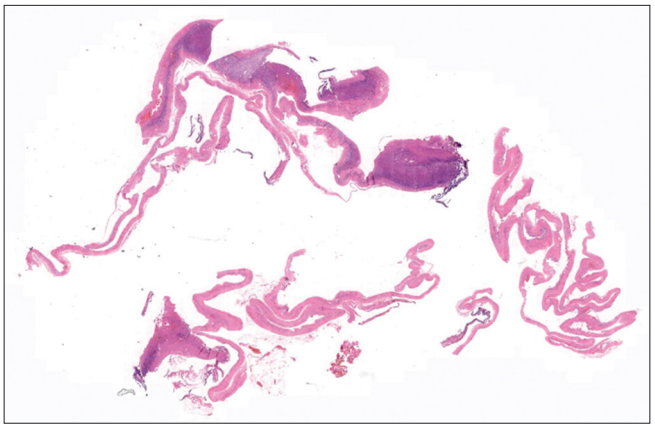

Figure 2: A 25-year-old male diagnosed with the left adrenal cyst. Section shows normal adrenal gland with fragments of the cyst wall.

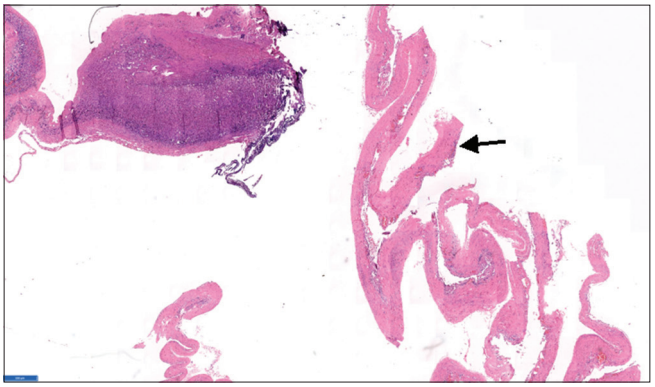

Figure 3: Section shows segment of cyst wall (Right, denoted by the arrowhead) with adjacent normal adrenal gland (Left)

Table 1: Previous cases with pathological findings.

Case	Clinical presentation	Pathological findings
1 (Subclinical Cushing's syndrome)	Incidental left adrenal mass in a 68-year-old Japanese woman, with obesity and hypertension	Vascular cyst with hyaline degeneration (lining cells were CD34 positive) within an adrenocortical adenoma. Immunohistochemical analysis of steroidogenic enzymes showed the expression of P450scc, 3βHSD, P450c17, and P450c21[1]
2 (Normal [nonfunctioning])	Incidental right renal and left adrenal tumors discovered in a 72-year-old Japanese female, with Type 2 diabetes mellitus	Right kidney: Renal clear cell carcinoma. Left adrenal: association of myelolipoma, endothelial cyst, and adrenocortical adenoma. Immunohistochemical analysis: Expression of cytochrome P450c17, 3H$^{\beta}$SD, and DHEA-sulfotransferase[2]
3 (Nonfunctioning adrenal mass)	Incidental right adrenal mass in a 53-year-old Chinese man with Type 2 diabetes	Cystic mass composed of fibrous wall tissues with local calcification. Another mass with a fibrous capsule outside the cystic wall composed of bright and dark cells, which are arranged in acinar and flaky shapes, and a large, deformed nucleus was present in the foci. Immunohistochemistry shows that the cells stained positive for CD34, D2-40, desmin, and SMA.[3]

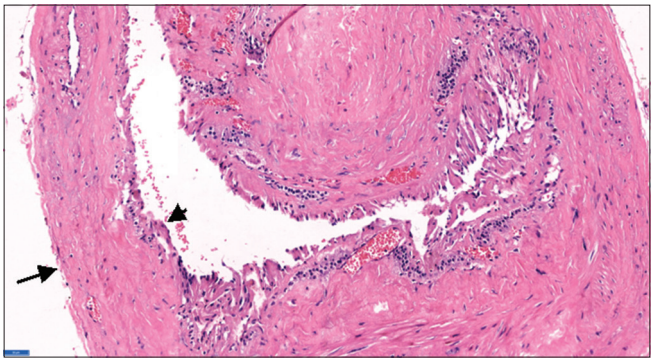

Figure 4: Section shows adrenal cyst wall lined by flattened epithelium (arrowheads) and congested blood vessels.

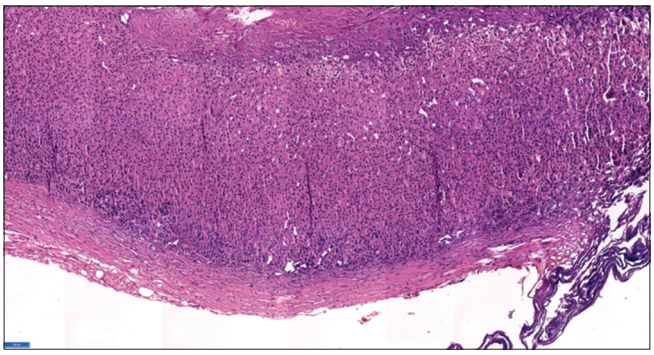

Figure 5: Section shows normal adrenal gland.

as lymphangiomatous cysts and angiomatous cysts. Routine hematoxylin and eosin staining will suffice for diagnosis, although the immunohistochemical markers would help in differentiating the lymphangiomatous subtype from the angiomatous subtype using podoplanin, a marker of lymphatic endothelium, which would be positive for CD31 and CD34 (Endothelial markers).

The pathogenesis of an adrenal endothelial cyst is a vastly underreported entity. While many have theorized that preexisting vascular lesions such as hamartomas, lymphangiectasis, or intraparenchymal hemorrhage may give rise to such cysts, our case showed no evidence of these findings, ruling out the same.[8-10]

At present, no protocols have been drafted concerning the management and treatment of adrenal cystic lesions, which can be attributed to their low incidence and difficulty in arriving to a conclusive pre-operative diagnosis. When malignancies are suspected clinically and an excess hormone secretion has been exhibited, the requirement of a multidisciplinary team becomes essential. Indications for surgical intervention in an adrenal cystic lesion include sizes larger than 5 cm due to the risk of hemorrhage and other complications, symptomatic presentations including, but not limited to endocrine abnormalities and malignancy. Other lesions which are asymptomatic and measure <5 cm can be adequately followed by imaging, although no screening algorithms have been drafted. In addition, fine-needle aspiration biopsy and cytological sampling of the cystic lesion may be performed in hormonally inactive cases, to specifically exclude pheochromocytoma in cases with suspicious imaging, as this pathology would radically alter the course of management. Laparoscopic adrenalectomy is still considered the gold standard in the management of adrenal cystic tumors.[3,13]

CONCLUSION

Adrenal cysts are rare clinical entities that commonly mimic more serious lesions such as adrenal malignancies and pheochromocytoma. They require thorough evaluation and histopathological examination to subclassify the cystic lesion and provide an appropriate diagnosis which is instrumental in instituting optimal treatment. While an

immunohistochemical workup may be used to confirm the diagnosis, morphology is the key to clinching the diagnosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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