Large adrenal cyst masquerading as a pancreatic cystic tumor: a rare diagnosis based on endoscopic ultrasound

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ABSTRACT
Adrenal cystic lesions constitute a rare finding and usually are diagnosed incidentally as retroperitoneal cysts during imaging studies. A major issue, especially for the large-sized ones, is their preoperative diagnosis and management, as imaging modalities often fail to detect their exact origin prior to surgery. We report a case of a big adrenal cyst that was mistakenly diagnosed and followed up as pancreatic cystic tail tumor. Our thorough workup, which included endoscopic ultrasound (EUS), managed to delineate an adrenal gland-originated cyst and altered further management of the patient. According to our knowledge, this is the first report in literature in which EUS proved its superiority over other imaging studies in clarifying the origin of an ambiguous big retroperitoneal cyst. We believe that for cases of large retroperitoneal cystic lesions, EUS consists the optimal diagnostic and sampling tool.

Introduction
Cystic lesions of the adrenal gland are infrequent entities. Few cases have been reported worldwide, only as case reports or series [1–3]. Adrenal cysts are usually an incidental finding and represent approximately less than 1% of incidentally discovered adrenal lesions [1]. Most of these lesions present no symptoms, unless they grow substantially in size (usually over 5 cm) or become complicated (hemorrhage, infection, or rupture). A major issue about their management is their preoperative diagnosis, as imaging modalities many times fail to determine their exact origin.

We report a case of a middle-aged patient with a retroperitoneal cystic lesion incidentally discovered in the past, which was presumably arising from the pancreatic tail according to previous imaging studies. She was inadequately followed up and referred to our department for a different cause. During a detailed workup, we confirmed the presence of the cystic lesion and performed endoscopic ultrasound (EUS) so as to further delineate the cyst. Our endosonographers managed to depict the cystic lesion and proved its origin from the adrenal gland.

Case report
A 47-year-old lady with symptoms of early satiety and flatulence referred to our department. She had a past medical history of arterial hypertension and mentioned the presence of a pancreatic cystic lesion incidentally diagnosed 9 years ago. The workup with abdominal Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) revealed a cyst containing solid components and measuring 11 × 8.4 cm, in close proximity to the pancreas and left kidney, presumably arising from the pancreatic tail (Fig. 1). The cyst had significantly grown in size compared to previous scanning. Carbohydrate antigen 19–9, carcinoembryonic antigen (CEA), and blood amylase were within normal limits. EUS was decided prior to surgery and was
performed with a curved line Olympus GF-UTC 180 echo-
endoscope. Surprisingly, EUS revealed a 12.5 cm × 8.5 cm well-
deﬁned cyst arising from the left adrenal gland (▶Fig. 2). We
could be conﬁdent that the lesion was originating from the
adrenal, as EUS enabled visualization of the pancreas and left
kidney and was clearly separated from them. It was conﬁned
by a thin wall and contained solid components and septa. We
shifted our processor (EU-ME2 Premier Plus, Olympus Medical
Systems, Tokyo, Japan) to harmonic software with a low me-
chanical index (0.15) and decided to proceed to contrast agent
administration (Sono-Vue 8 mL/vial, Bracco, Amsterdam, Hol-
land). We injected the microbubble agent as an intravenous bo-
lus from a peripheral vein and observed the lesion’s enhance-
ment behavior for 120 seconds. We noticed that the cyst did
not uptake contrast except for its wall. Aspiration cytology a-
analysis revealed sparse mast cells and lymphocytes but no ma-
lignancy. Cystic ﬂuid was serous, with CEA and amylase levels
within normal limits. We decided to verify the functional status
of the adrenal lesion preoperatively. The patient was referred to
endocrinologist and biochemical assessment with 24-h urine
metanephrines, catecholamines, aldosterone, 17 hydrocorti-
costeroids, and 17 ketokosteroids was performed. Moreover,
blood ACTH (adrenocorticotropic hormone), DEAS (dehydroe-
piandrosterone), aldosterone, cortisol, and plasma rennin ac-
tivity levels were calculated and all were within normal limits.
The patient was referred to our multidisciplinary meeting in
which surgery was decided. During operation and due to signif-
ican t encountered adhesions, the lesion could not be easily re-
sected and the patient needed to undergo peripheral pancrea-
tectomy, splenectomy, left adrenalectomy, nephrectomy, and
segmental colectomy. Histopathologically, the wall of the re-
sected cyst composed of ﬁbrous tissue along with entrapped
adrenal cortical and medullary cells, suggesting the diagnosis
of an adrenal cyst. Differential diagnosis included a pseudocyst
with hemorrhagic content or an endothelial cyst of lymphatic
origin, since they both share common histological ﬁndings.

Discussion

Adrenal cysts are very rare entities, mainly diagnosed in autops-
y studies in 0.06–0.18% of the population [1, 2]. They are clas-
siﬁed into 4 categories: pseudocysts, endothelial cysts, infe-
tious cysts, and solid tumors with cystic degeneration. These
cysts usually remain asymptomatic, unless they become secre-
tory, get complicated, or produce symptoms from pressure to
adjacent structures due to their growing size. Most commonly,
these cysts present in the fourth or ﬁfth decade of life, as it was
the case for our patient as well. The incidence of malignancy
in adrenal cystic lesions is less than 7% and this is largely related
to their size, as adrenal cysts over 6 cm can carry an increased
risk for cancer [4]. Surgical treatment is strongly suggested for
cysts larger than 5 cm or for complicated ones [5].

Physicians should always keep in mind the adrenal-origina-
ted tumors in the differential diagnosis of retroperitoneal le-
sions. We report a case of a massive retroperitoneal cyst that
was initially misdiagnosed as pancreatic cystic tumor. Despite
improvements on imaging, the origin of retroperitoneal tu-
mors, especially the sizable ones, may remain ambiguous.
Usually, the initial imaging workup includes transabdominal ul-
trasound, CT and MRI, which in many cases fail to detect the ex-
act cystic origin and the patient is led misdiagnosed to the sur-
gical bed [4–6]. This can have an impact on the surgical proce-
dure and to the patient’s clinical outcome as well. A rare case of
a large cystic pheochromocytoma was reported lately, where
EUS failed to determine its origin and was ﬁnally diagnosed in-
traoperatively [6]. Another interesting case was that of a giant
hemorrhagic cyst, where only contrast enhanced transabdomi-
nal ultrasonography managed to determine the exact origin of
the lesion, compared to the inconclusive diagnosis of conven-
tional ultrasound and CT [4].

Another clinically signiﬁcant issue is that of the preoperative
functional activity of adrenal lesions. Usually, solid masses
should be examined for secretory activity with ACTH, aldosterone, androgens, metanephrines, and cortisol products (blood and urines). Cystic masses very rarely present any secretory activity, but large cystic masses when diagnosed as adrenal originated ones should always be investigated for hormone production prior to surgery and especially if they present solid components [7].

**Conclusion**

Retroperitoneal adrenal cystic lesions and especially the large ones are sometimes very difficult to be accurately diagnosed preoperatively. EUS is an optimal method for depiction of the adrenal gland and whenever available it could be a first-line imaging modality for the evaluation of retroperitoneal cysts. This is the first report in the literature that EUS proved to be superior to all other imaging studies for preoperative diagnose of an adrenal cystic lesion.

**Competing interests**

None

**References**

[1] 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

[2] Bellantone R, Ferrante A, Raffaelli M et al. Adrenal cystic lesions: report of 12 surgically treated cases and review of the literature. J Endocrinol Invest 1998; 21: 109 – 114

[3] Chodisetti S, Boddepalli Y, Kota M. Giant adrenal cyst displacing the right kidney. Indian J Urol 2016; 32: 81 – 82

[4] Cantisani V, Petramala L, Ricci P et al. A giant hemorrhagic adrenal pseudocyst: contrast-enhanced examination (CEUS) and computed tomography (CT) features. Eur Rev Med Pharmacol Sci 2013; 17: 2546 – 2550

[5] Lal TG, Kaulback KR, Bombonati A et al. Surgical management of adrenal cysts. Am Surg 2003; 69: 812 – 814

[6] Lee J, Raman K, Sachithanandan S. Cystic pheochromocytoma mimicking a malignant pancreatic cyst. Gastrointest Endosc 2011; 74: 712 – 713

[7] Sioka E, Symeonidis D, Chatzinikolaou I et al. A giant adrenal cyst difficult to diagnose except by surgery. Int J Surg Case Rep 2011; 2: 232 – 234