CASE SERIES

Case series on Cystic Adrenal Lesions: A single Institution experience

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Abstract:

Adrenal cysts are rare and uncommon disease with only around 600 cases reported so far. They are usually asymptomatic or may rarely present with abdominal pain or fullness. Different types of adrenal cyst have been described. They are usually benign in nature. Optimum management of adrenal cysts still remain controversy, owing to its low incidence. We report four cases with different histological types of adrenal cysts, their manifestations and management.

Keywords: Adrenalectomy, adrenal cyst, retroperitoneal, laparoscopic

Introduction:

Adrenal cysts are rare and uncommon, with a reported incidence of 0.064% to 0.18%in autopsy studies [1]They account for 4-22% of all adrenal incidentaloma [2-4]Although more common in 3rd-6th decades, they can present in any age, with a female preponderance.[5] Only few case reports and case series has been published so far. Owing to its rarity, there is no specific guideline on the management of adrenal cyst so far. Herein, we report 4 patients of adrenal cysts, one each of pseudocyst, true cyst and endothelial cyst and rare case of adrenocortical carcinoma arising from a pseudocyst.

Patient 1:

A 35-year-old lady who was under investigation for hepatic cystic lesion was referred to our department for further evaluation. She had initially presented with complaints of pain in right upper abdomen for five months which was insidious in onset, dull aching, non-radiating and was relieved only on medication. There was no abdominal distension, vomiting, altered bowel or bladder habits. She had no history suggestive of a functional adrenal mass. The patient had no co-morbidities or significant past history. Her general and abdominal examination were essentially normal. Biochemical investigations showed a normal liver, renal and adrenal functioning. ELISA for Echinococcus was negative.

Ultrasonography of the abdomen, which was performed before the referral revealed an irregular cystic shadow, measuring 35x33mm in the posterior segment of right lobe of liver with foci of calcification, suggestive of right hepatic hydatid cyst. [Fig 1a]

CECT of the abdomen revealed a multiloculated cystic lesion measuring 31x32x36mm with multiple wall and septal calcification in right adrenal region. Right adrenal gland was not visualised separately from the lesion. The image was suggestive of an adrenal cystic lesion. [Fig 1b]

In view of patient's symptoms, surgical management was planned and retroperitoneoscopic adrenalectomy was performed. Intraoperatively, the lesion appeared multiloculated containing straw coloured fluid. It measured 4x3 cm in size, with all borders free. [Fig 1c] Both intraoperative and post-operative periods were uneventful and the patient was discharged on post-operative day 2. The post-operative histopathology showed a cystic lesion with fibro collagenous wall and no evident lining. Remnant of adrenal tissue is seen at the outer aspect. Histology was consistent with Adrenal Pseudocyst. [Fig 1d]

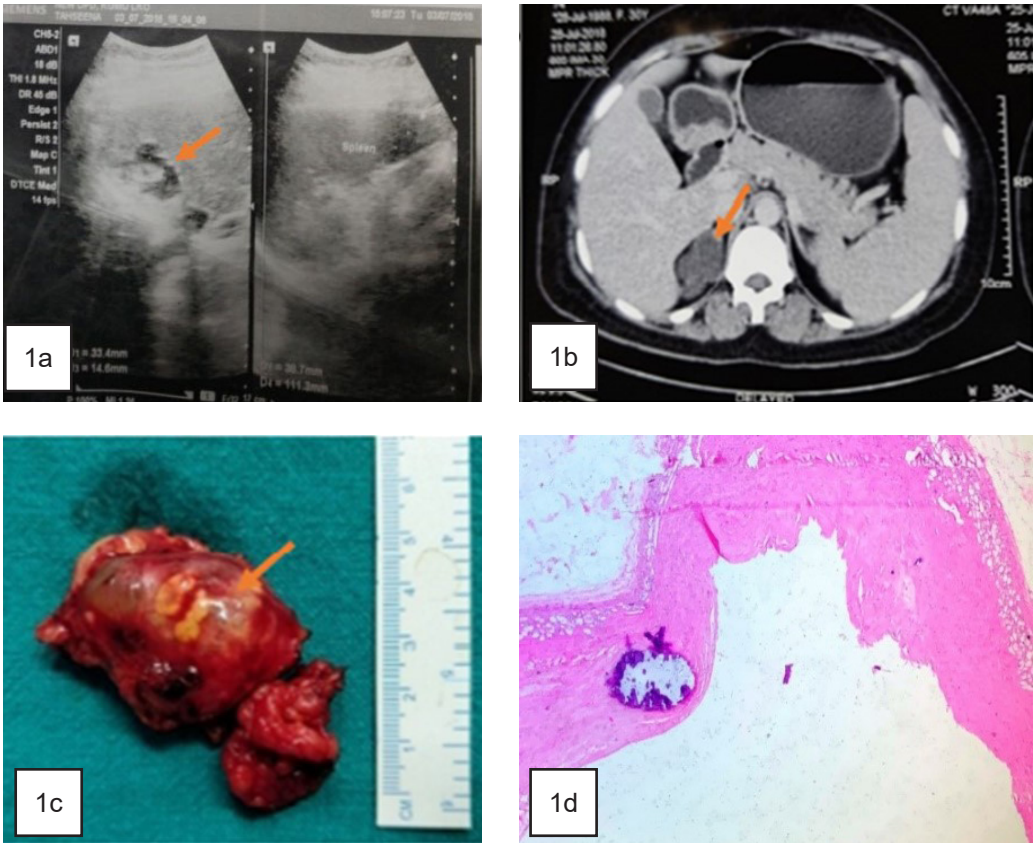


Figure 1: Adrenal pseudo cyst. a) CECT showing a multiloculated cystic lesion measuring 31x32x36 mm with multiple wall and septal calcification in right adrenal region (arrow); b) Resected specimen showing multi loculated lesion filled with fluid and; c) Histopathology shows fibro collagenous cyst wall with focal calcification and no lining. Remnant of adrenal tissue is seen at the outer aspect (Hematoxylin and eosin; 400 X).

Patient 2:

A 29 years old lady presented with complaints of dull aching pain in right hypochondriun, radiating to back. She had similar episode 7 and 3 years back for which she underwent two times guided aspiration of right suprarenal cyst. She also had history of image guided pigtail insertion for the same done elsewhere, 3 months back which was removed after 8 days. She had no history suggestive of a functional adrenal mass. Her general and per abdominal examination were essentially normal. Biochemical investigations showed a normal liver, renal and adrenal functioning.

USG abdomen revealed a mixed echogenic mass in right suprarenal region containing thick fluid (vol-449cc). [Fig 2a]

CECT abdomen revealed a well-defined rounded hypodense lesion in region of the right adrenal gland measuring approx. 79x75x78 mm with slight hyperdense echogenic area and presence of tiny specks of calcification in peripheral part with no fat component or enhancing nodule. [Fig 2b]

In view of patient history of multiple aspiration and guided drain insertion, she was managed with open adrenalectomy via trans-peritoneal anterior approach. Intra-operatively there was 8x7 cm solid cystic mass containing haemorrhagic fluid. [Fig 2c] Her histopathology revealed fibrocollagenous cyst wall lined by flattened endothelial cells along with remnants of adrenocortical tissue consistent with endothelial cyst. [Fig 2d]

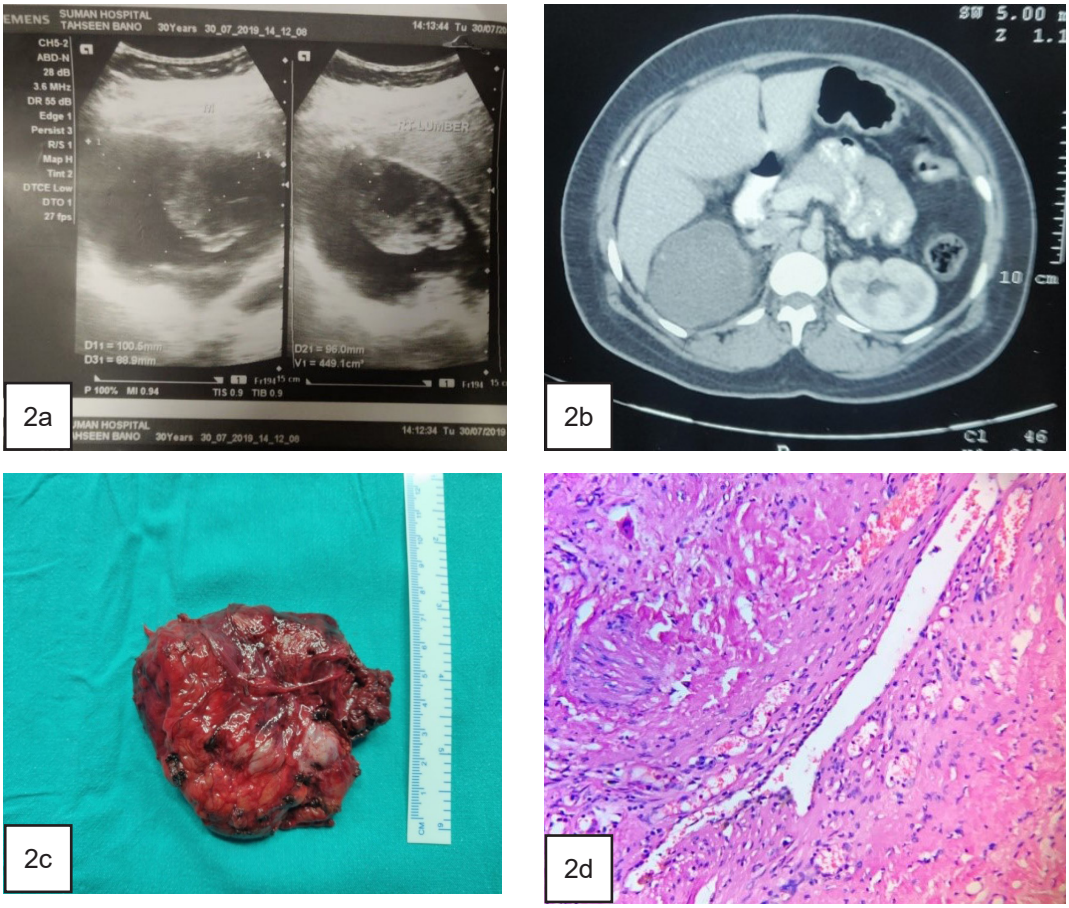


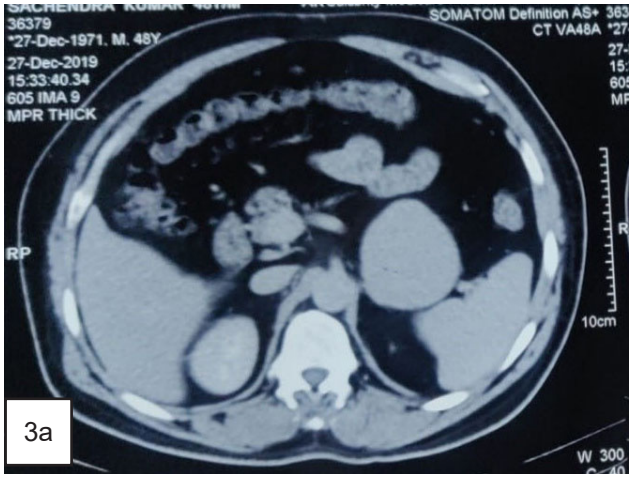
Figure 2: Adrenal Endothelial cyst. a) USG abdomen showing a mixed echogenic mass in right suprarenal region (arrow); b) CECT abdomen showed well defined right adrenal cyst as round hypo dense lesion measuring approx. 79x75x78 mm with internal haemorrhage as slight hyper dense echogenic area in it (arrow); c) Resected specimen showing right suprarenal mass measuring 8 x 7 cm with cystic and solid component and; d) Histopathological evaluation display cystic structure with fibrocollagenous wall lined by flattened endothelial cells and red blood cells in the lumen (Hematoxylin and eosin; 400 X).

Patient 3:

A 48-years aged gentleman who was under evaluation for abdominal fullness and dyspepsia from Medical Gastroenterology was diagnosed to have left adrenal lesion for which he was referred to our department for further evaluation. He had no history suggestive of a functional adrenal mass. The patient had no co-morbidities or significant past history. His general examination and abdominal examination were essentially normal. Biochemical parameters including hemogram, liver, renal and adrenal function were within normal limits.

CECT abdomen showed well-defined mildly hyperdense thin walled non enhancing lesion with HU 95 measuring 6.3x6.4x6.6 cm in the retroperitoneum closely abutting pancreas and left adrenal with subtle calcification at inferior aspect of lesion? Adrenal cyst. [Fig 3a]

Although the patient had no significant symptoms, since the size of the lesion was > 6cm, surgery was planned and he was managed by laparoscopic adrenalectomy via anterior approach. Intraoperatively, there was a well circumscribed cystic mass lesion measuring 6 x 5 cm. His final HPE was consistent with true adrenal cyst. [Fig 3b]



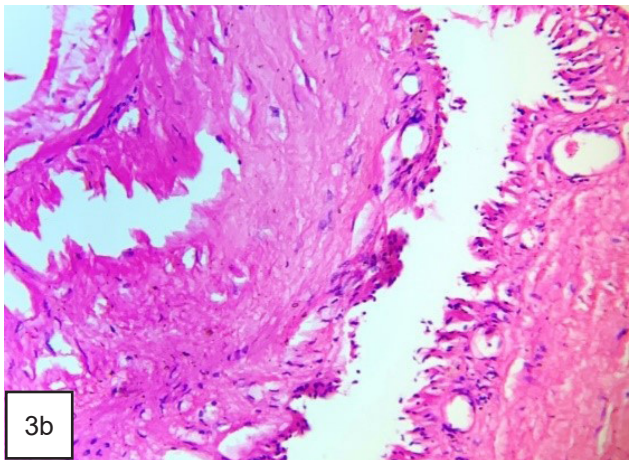


Figure 3: Epithelial adrenal cyst. a) USG abdomen showing (arrow) a well-defined hypo-echoic lesion in left suprarenal region measuring 64x52 mm (? adrenal origin); b) CECT showing a well define, round to oval, hypo dense lesion with hyper dense thin wall measuring 6.3 x 6.4 x 6.6 cm in the retro peritoneum closely abutting pancreas (arrow); c) Cyst wall lined by single layer of columnar epithelial cells (Hematoxylin and eosin; 400 X).

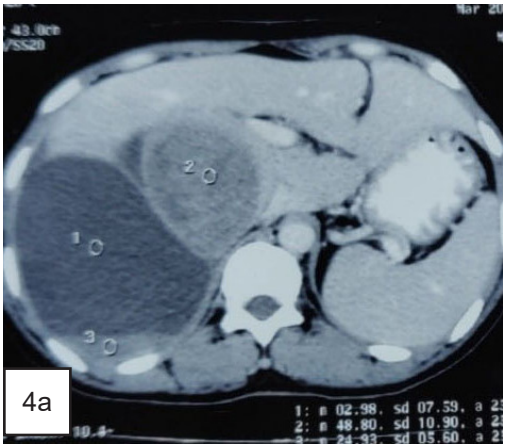
Patient 4:

A 40 years aged lady, presented with pain in right hypochondrium and lumbar region for 1 year which was associated with generalised weakness, fatigue and constipation for the past one month. No history suggestive of a functional adrenal mass. On per-abdominal examination, a lump of size 13x10 cm was palpable in right hypochondrium extending to lumbar region, crossing the midline which was firm to hard in consistency with smooth surface. Serum and urinary catecholamines were unremarkable. During investigation, a well-defined para-renal 13×10 cm septated cyst was identified on CT. The right adrenal gland was not separately identified from the mass, suggesting adrenal origin. [Fig4a]

In view of large size and malignant potential of complex cysts, right open adrenalectomy was performed via anterior transperitoneal approach. Per-operatively, there was a very large predominantly cystic mass measuring 15x15cms with solid areas which was densely adhered to inferior surface of liver.

Histopathological evaluation revealed a cystic lesion with fibrocollegenous cyst wall and absence of any lining. These were a solid component composed of sheets of atypical cells with resemblance of cortical cells. [Fig 4b] Mitosis was >5/10 high power field. Areas of necrosis and haemorrhage were also seen along with presence of atypical mitotic figures. The modified Weiss score was 4 suggesting

adrenocrtical malignancy. These tumour cells were immunohistochemically positive for synaptophysin, vimentin, melan A and inhibin with no expression of chromogranin and CK7, CK20. Ki67 proliferation index was 5%. Hence, the case was finally diagnosed as Adrenocortical carcinoma arising from a pseudocyst with poor prognostic markers. The patient is lost to follow up.



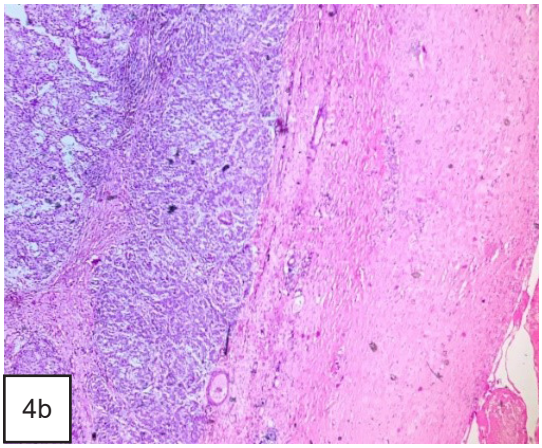


Figure 4a: Cystic adrenocortical carcinoma. a) CT scan shows well-defined para-renal 13×10 cms septated cyst (arrow) compressing the adjacent bowel and right lobe of liver. The right adrenal gland was not separately identified from the mass, suggesting adrenal origin and b) Microscopy shows fibrocollagenous cyst wall with sheets of atypical epithelial cells having high nucleo-cytoplasmic ratio, pleomorphic nuclei and moderate amount of eosinophilic cytoplasm (Hematoxylin and eosin; 400 X).

Discussion

Adrenal cyst was first described by Viennese Anatomist Greiselius in 1670 in a 45 year old man who died due to rupture of an adrenal cyst weighing more than 4 kg.[6] The reported female to male ratio in literature is 3:1. [5] They are usually unilateral and bilateral cysts are seen in only about 8-15 % of cases .[1] Most of these cysts are benign in nature with reported incidence of malignancy in only 7 % of cases.[7]About 95% of these malignant lesions are metastases from other primary epithelial tumours(lung, kidney, colon, breast, pancreas, liver and stomach), 3% are pheochromocytoma and remaining 2% adrenocortical carcinomas.[3, 8] They are usually asymptomatic and are discovered incidentally. However, in about 39% of cases, they may present with large mass lesions and pain due to haemorrhage or cyst rupture. Rarely (9% of cases) adrenal cysts are associated with hypertension, probably due to compression of adrenal artery or renal medulla [3]. In our series, three of them had different modes of presentation. Our first patient had small but symptomatic adrenal mass, second patient presented with history of multiple aspirations and the third patient presented with large adrenal mass with complaints of only vague abdominal fullness.

The first classification of adrenal cyst was given by Terrier and Lecene in 1906. They classified adrenal cyst into haemorrhagic, endothelial, congenital retention, cystic adenomas and parasitic types.[9] Following which many other classifications were formulated eventually. However, the most accepted classification till date was given by Foster in 1996. He classified adrenal cyst into four types based on histological types on autopsy and incidence: Endothelial cyst (45%), pseudocyst (39%), epithelial cyst (9%), and parasitic cyst (7%).[6]

Pseudocysts are most common among all adrenal cysts across different studies with an incidence of 39%. [6] They are usually large and uniloculated with walls devoid of any cellular lining.They vary greatly in size, ranging from few millimetres to more than 50cm.[11] Association of pseudocyst with adrenal neoplasm has been reported in about 18.7-44% of cases.[12] Among the malignancies found in adrenal pseudo cysts; adreno-cortical carcinoma (ACC) is by far the most common [13].

Endothelial cyst are also known as simple cysts. They are the most common among adrenal cysts in autopsy series with incidence of 45%, but account for only 2-24% of clinically symptomatic lesions. They are usually small in size; with an average size of less than 2cm.The walls of these cysts are lined by

smooth flattened endothelial lining [7]. Two subtypes of endothelial cyst are described; lymphangiomatous type (94%) and angiomatous type [6%].[5, 7]Epithelial cyst are true cysts. They are mesothelial in origin and their walls are lined by smooth flattened epithelial lining [12]. Different subtypes of epithelial cysts are reported, namely glandular or retention cyst, cystic adenomas, and embryonal cyst.[12] Parasitic cyst are rare with an incidence of 7 % [6]. Echinococcosis is the most common causative organism. They have thick walls with or without calcification.

Till date there have been only around 600 cases of adrenal cyst reported so far. The largest case series to date was reported by Erickson LA et al, in 2004, where he reviewed 41 cases of adrenal cyst (32 pseudocyst, 8 endothelial cyst and 1 epithelial cyst). [10]

Imaging modalities for adrenal cyst include ultrasound abdomen, which has a reported sensitivity of 60-70% for detecting adrenal cyst.[11] Adrenal cyst on ultrasound reveal well-defined, round to oval anechoic structure showing posterior acoustic enhancement. Hyperechoic pattern may also be seen on ultrasound in case of haemorrhage in the cyst. Contrast enhanced computed tomography [CECT] of the abdomen is the gold standard imaging modality with a sensitivity of 85-100% and specificity of 95-100% [11]. On CECT, true cysts characteristically have fluid attenuation, usually less than 20 HU, have smooth borders with thin non-enhancing walls. Lack of contrast enhancement on CT favours the diagnosis of adrenal cyst. Calcification can be noted in around 15-70% of cases which can be either rim or nodular calcification. MRI has a sensitivity of 100 %.[15] On MRI, simple cyst appear hypo intense on T1 weighted images and hyper intense on T2 weighted images without any soft tissue component or internal enhancement. In case of haemorrhage in adrenal pseudocysts, they appear hyper intense on both T1 and T2 weighted MRI images.

Optimum management of adrenal cysts still remain a controversy, owing to its low incidence. Surgical management, whether open or minimally invasive depends on a surgeon's preference, adrenal lesion size and characteristics on imaging. Surgery is usually indicated in functional cysts, malignant or potentially malignant cysts, symptomatic cysts of any size, asymptomatic cysts of size more than 5cm and those patients with uncertain follow up.[7,16]

In our first patient, surgical removal was done in spite of lesion being <4 cm in greatest dimension, due to presence of chronic pain. The remaining cases also underwent adrenalectomy because of larger size as well as presence of symptoms. For lesions measuring more than 6 cm, open adrenalectomy was found to be a better operative approach, as these lesions were found to have dense adhesions with adjacent structures (seen in 3 out of 4 patients). The reason for this is not clear, but it may be due to cystic fluid permeating through the capsule causing adhesions with adjacent tissues. We also experienced this adhesive plane making difficult dissection in our patient series.

Chien et al, in 2008 reviewed the importance of surgical management in patients with adrenal cyst wherein he reported 25 cases of adrenal cyst (16 pseudocyst, 8 endothelial and 1 epithelial cyst) where seven adrenal pseudocyst were associated with tumour including two pheochromocytomas, one neuroblastoma, one adrenal cortical carcinoma, one adrenal cortical adenoma, one myelolipoma, and one schwannoma. He concluded that because of their heterogeneous aetiology and overlapping clinical findings, definite diagnosis relies on extensive sampling and thorough microscopic examination in order to exclude the possibility for coexisting tumour.[12]

Conservative management is apt in those with uncomplicated/asymptomatic cysts <5cm.[12]A minimum of 18 months of follow up with repeat CT every 6 months is indicated. Aspiration of cyst can be considered as an alternative to surgery in case of surgically unfit patients.[1, 5, and 7] Marsupialisation or decortication have also been tried as alternatives to surgery for large cyst specially those cyst which are adherent to multiple organs where excision may be difficult[5]. Sclerotherapy using absolute alcohol has also been described but it is associated with high recurrence of 30-50 % [1, 5].

In conclusion cystic adrenal lesions are rare and uncommon disease with varied manifestations and sometime present with diagnostic dilemma. Proper investigation including CT or MRI is essential for defining adrenal cystic lesion and also for differentiating it from cystic lesion of adjacent organs. Surgery is the treatment of choice in symptomatic case and histopathological examination is essential for definitive diagnosis.

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