

## CASE REPORT

# All Right Upper Abdominal Cysts are Not Hepatic Cyst

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## ABSTRACT

Adrenal cysts are rare and uncommon disease with only around 600 cases reported so far. Cystic lesion in right hypochondria is usually of hepatic in origin. The right adrenal cyst may sometimes be confused as the hepatic hydatid cyst. We herein report the case of a patient with the adrenal pseudocyst who initially presented with features of the hepatic hydatid cyst.

**Keywords:** Adrenal cyst, Adrenalectomy, Hydatid cyst, Retroperitoneal.

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## BACKGROUND

Cystic lesions in right hypochondrium are usually of the hepatic origin. Rarely, cystic lesions from adjacent organs can mimic hepatic cysts. Adrenal cysts are rare and uncommon, with a reported incidence of 0.064–0.18% in autopsy studies.<sup>1</sup> They account for 4–22% of all adrenal incidentaloma.<sup>2–4</sup> Although more common in third to sixth decades, they can present in any age, with a female preponderance.<sup>5</sup> We report an interesting case of a woman who initially presented with features of the hepatic hydatid cyst, further investigations suggesting an adrenal incidentaloma and postoperative histopathology revealing an adrenal pseudocyst.

## CASE DESCRIPTION

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 the right upper abdomen for 5 months, which was insidious in onset, dull aching, nonradiating, 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 comorbidities or significant past history. Her general examination 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 35 × 33 mm in the posterior segment of the right lobe of the liver with foci of calcification, was suggestive of the right hepatic hydatid cyst (Fig. 1).

Contrast-enhanced computed tomography (CECT) of the abdomen revealed a multiloculated cystic lesion measuring 31 × 32 × 36 mm with multiple wall and septal calcification in the right adrenal region. The right adrenal gland was not visualized separately from the lesion. The image was suggestive of an adrenal cystic lesion (Fig. 2).

In view of patient's symptoms, surgical management was planned and retroperitoneoscopic adrenalectomy was performed. Intraoperatively, the lesion appeared multiloculated containing straw-colored fluid. It measured 4 × 3 cm in size, with all borders free (Fig. 3). Both intraoperative and postoperative periods were uneventful and the patient was discharged on postoperative day 2. The postoperative histopathology showed a cystic lesion with

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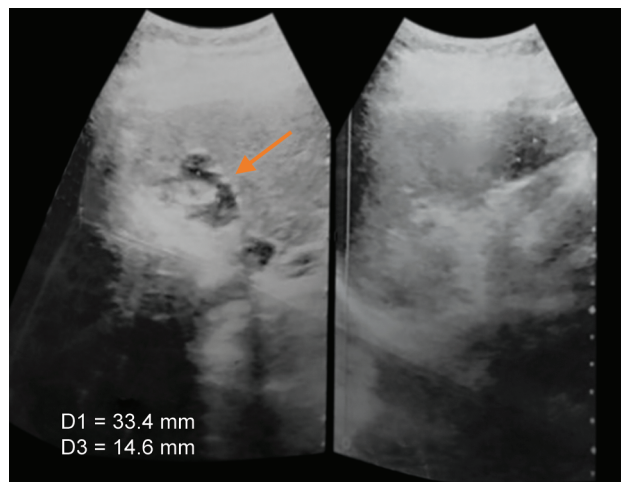
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fibrocollagenous wall and no evident lining. Remnant of adrenal tissue is seen at the outer aspect. Histology was consistent with adrenal pseudocyst (Fig. 4).

## DISCUSSION

The 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.<sup>6</sup> The reported female to male ratio in literature is 3:1.<sup>5</sup> They are usually unilateral and bilateral



**Fig. 1:** Ultrasound image of right adrenal cyst confused as hepatic cyst

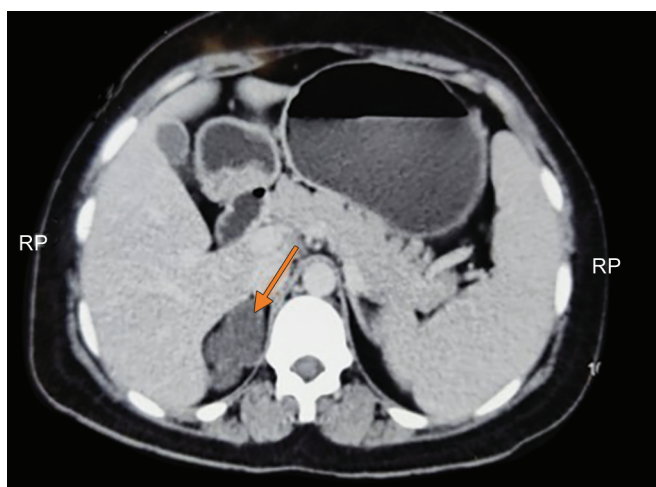


Fig. 2: CECT abdomen image confirming right adrenal cyst



Fig. 3: Operative specimen of right adrenal cystic mass

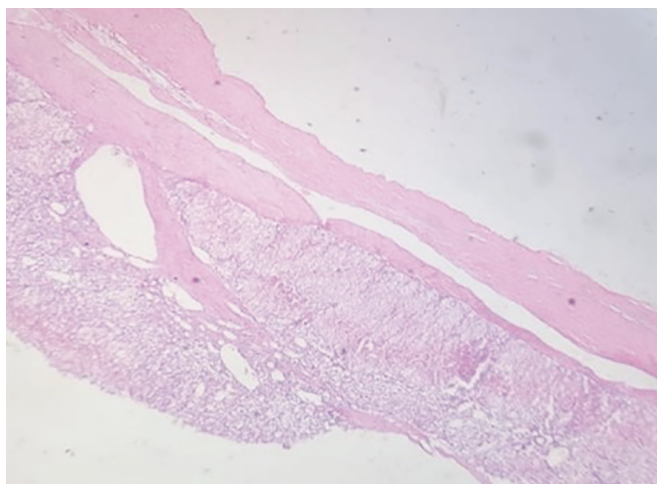


Fig. 4: Histopathology image of right adrenal pseudocyst

cysts and are seen in only about 8–15% of cases.<sup>1</sup> Most of these cysts are benign in nature with reported incidence of malignancy in only 7% of cases.<sup>7</sup> About 95% of these malignant lesions are metastases from other primary epithelial tumors (lung, kidney, colon, breast, pancreas, liver, and stomach), 3% are pheochromocytoma, and remaining 2% are adrenocortical carcinomas.<sup>3,8</sup> 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 hemorrhage and cyst rupture. Rarely (9% of cases), adrenal cysts are associated with hypertension, probably due to compression of the adrenal artery or renal medulla.<sup>3</sup>

Terrier and Lecene in 1906 first classified adrenal cysts into hemorrhagic, endothelial, congenital retention, cystic adenomas, and parasitic types.<sup>9</sup> Many other classifications were formulated eventually. In 1966, Foster classified the adrenal cyst into four types based on histological types and incidence: endothelial cyst (45%), pseudocyst (39%), epithelial cyst (9%), and parasitic cyst (7%). This remains the most accepted classification till date.<sup>6</sup>

### Pseudocyst

Pseudocysts are most common among all adrenal cysts across different studies with an incidence of 39%.<sup>6</sup> They are usually large

and uniloculated with walls composed of the fibrous connective tissue devoid of any cellular lining. The walls are usually 1–5 mm thick, rarely reaching up to 3 cm.<sup>6</sup> Islands of the adrenal cortical tissue may be seen incorporated into the walls of the tumor in about 19% patients.<sup>10</sup> They vary greatly in size, ranging from few millimeters to more than 50 cm.<sup>11</sup> Association of pseudocyst with adrenal neoplasm has been reported in about 18.7–44% of cases.<sup>10</sup> Adrenal cysts can also be associated with variety of other conditions like polycystic renal disease,<sup>12</sup> Beckwith-Wiedemann syndrome,<sup>13</sup> Klippel-Trenaunay-Weber syndrome,<sup>14</sup> abdominal aneurysm, and schwannomas.<sup>15,16</sup> Rarely, they have been found in association with pregnancy.<sup>17</sup> The etiopathogenesis includes hemorrhage within normal adrenal tissue or tumor, cystic degeneration of primary adrenal neoplasm, vascular neoplasm, and malformation. They may also result from other pathological conditions like toxemia of pregnancy, diabetes, uremia, toxemia of disseminated meningococcal infection, Waterhouse Friderichsen syndrome, acute crush injuries, and thrombosis of adrenal veins.<sup>6</sup> Few researchers have reported an apparent vascular origin for pseudocysts, which is substantiated by the positive staining for endothelial markers in the walls of tumors.<sup>18</sup>

### Endothelial Cyst

They 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.<sup>7</sup> They are usually small in size, with an average size of less than 2 cm. Bilateral lesions can be seen in about 8% of cases.<sup>1</sup> The walls of these cysts are lined by a smooth flattened endothelial lining.<sup>7</sup> Two subtypes of the endothelial cyst are described: lymph angiomatous type (94%), which arise from ectasia of lymphatic vessels; and angiomatous type [6%] arising from ectatic blood vessels.<sup>5,7</sup> Immunohistochemically, endothelial cysts express CD31 and factor VIII antigen.<sup>5,7</sup>

### Epithelial Cyst

These are true cysts, with an incidence of 6–9%.<sup>6</sup> They are mesothelial in origin and their walls are lined by smooth flattened epithelial lining.<sup>19</sup> Different subtypes of epithelial cysts are reported, namely glandular or retention cyst, cystic adenomas, and embryonal cyst.<sup>10</sup> Immunohistochemical staining of the walls demonstrates antibodies to keratin AE1/AE3.<sup>20</sup>

## Parasitic Cyst

These are rare with an incidence of 7%.<sup>6</sup> Echinococcosis is the most common causative organism. They have thick walls with or without calcification. Eosinophilia can be noted in 20% of cases and are serologically positive in 90% of cases.<sup>7</sup>

As far as imaging is concerned, ultrasound has a reported sensitivity of 60–70% for detecting the adrenal cyst.<sup>21</sup> The adrenal cyst on ultrasound reveals well-defined, round to oval anechoic structure showing posterior acoustic enhancement. The hyperechoic pattern may also be seen on ultrasound in case of hemorrhage in the cyst. Contrast-enhanced computed tomography of the abdomen is the gold standard imaging modality with a sensitivity of 85–100% and specificity of 95–100%.<sup>21</sup> On CECT, true cysts characteristically have fluid attenuation, usually lower than 20 HU, have smooth borders with thin nonenhancing walls. Lack of contrast enhancement on CT favors the diagnosis of the 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%.<sup>21</sup> On MRI, simple cyst appears hypointense on T1-weighted images and hyperintense on T2-weighted images without any soft tissue component or internal enhancement. In case of hemorrhage in adrenal pseudocysts, they appear hyperintense on both T1- and T2-weighted MRI images.

Optimum management of adrenal cysts still remains a controversy, owing to its low incidence. Surgical management, whether open or minimally invasive, depends on a surgeon's preference and expertise. Surgery is indicated in functional cysts, malignant or potentially malignant cysts, symptomatic cysts of any size, asymptomatic cysts of size more than 5 cm, and those patients with uncertain follow-up.<sup>7,22</sup> Conservative management is apt in those with uncomplicated/asymptomatic cysts <5 cm.<sup>22</sup> 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.<sup>1,5,7</sup> Marsupialization or decortication has also been tried as alternatives to surgery for large cysts especially those cysts that are adherent to multiple organs where excision may be difficult.<sup>5</sup> Sclerotherapy using absolute alcohol has also been described but it is associated with high recurrence of 30–50%.<sup>1,5</sup>

## CONCLUSION

In conclusion, cystic adrenal lesions are rare and uncommon disease and may present as diagnostic dilemma. Proper investigation including CT or MRI is essential for defining adrenal cystic lesion and differentiating 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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