Giant Adrenal Cyst : Epithelial Variant, A Rare Entity

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ABSTRACT

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Adrenal cystic lesions are a rare entity encountered in clinical practice. They can be classified into 4 types – endothelial, pseudocyst, epithelial and parasitic. It is mostly asymptomatic and non-functional. Lack of definite laboratory and radiological diagnostic criteria makes it difficult to establish their diagnosis preoperatively and excision is needed in most cases. Laparoscopic adrenalectomy is the treatment of choice although small, benign, asymptomatic, non functional lesions can be aspirated or put on surveillance. Epithelial cyst is one of the rare types of adrenal cyst comprising only 9% of all adrenal cysts with most of the benign cysts less than 7 cm. Hereby illustrating a rare case of incidentally diagnosed giant epithelial adrenal cyst, the incidence of which has been less than hundred in the literature reported so far

Keywords: Epithelial Variant, Giant Cyst, Adrenal Cyst in Pregnancy, Adrenalectomy

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INTRODUCTION

Adrenal cysts are a well known entity with a documented incidence of 0.064% to 0.18% in autopsy series. The endothelial and pseudocyst variants of adrenal cysts are most common with epithelial subtype contributing to only 9% of all adrenal cysts. Given the chance of associated malignancy, the cysts need to be kept under close follow up or removed surgically. Also the absence of definitive diagnostic criteria demands excision of the cyst for accurate histopathological diagnosis given the close resemblance of the cysts to adrenal cystic neoplasms.

CASE REPORT

A 27 year old female patient, during routine antenatal evaluation, was found to have a right upper abdominal space occupying lesion. 3 months post partum, she was referred for detailed evaluation of the mass lesion. On history, she never had any complaints of pain abdomen, hematuria, dysuria, LUTS, fever, vomiting, early satiety, breathing difficulty, loss of weight or appetite, jaundice, headache, seizures, bone pain. She was clinically asymptomatic. She was a known case of hyperthyroidism on medication and had history of hypertension during pregnancy. Antenatal period was otherwise uneventful and she underwent elective LSCS in view of high risk due to the mass. On examination, her BMI was 28, no pallor or pedal oedema. She was normotensive and per abdomen examination showed a soft cystic swelling in the right upper abdomen with rounder borders, not moving with respiration which was mildly tender. A healed pfannenstiel LSCS scar was also visible. Blood parameters were within normal limits. On evaluation for adrenal mass, urinary VMA was negative and serum cortisol was within normal limits.

On radiological evaluation, ultrasound abdomen showed a large 18x13 cm right hypochondrial hypoechoic cystic lesion. MRI abdomen with contrast was done which showed a 13.5 x 11.4 x 11.9 cm uniloculated oval to round shaped thin walled retroperitoneal cystic lesion with equivocal wall enhancement causing splaying of medial and lateral limbs of right adrenal gland – most likely to be an adrenal cyst **(figure 1).**

In view of the large swelling, patient underwent open right adrenalectomy. Intraoperatively a tense cystic swelling was seen between liver and upper pole of right kidney arising from the right adrenal gland, with adrenal tissue seen above and below the swelling. The cyst contained yellow clear fluid which was aspirated and sent for analysis. The cyst wall along with the right adrenal gland was excised. Post operative period was uneventful and the patient was discharged on POD 6.

Histological examination showed a uniloculated cyst

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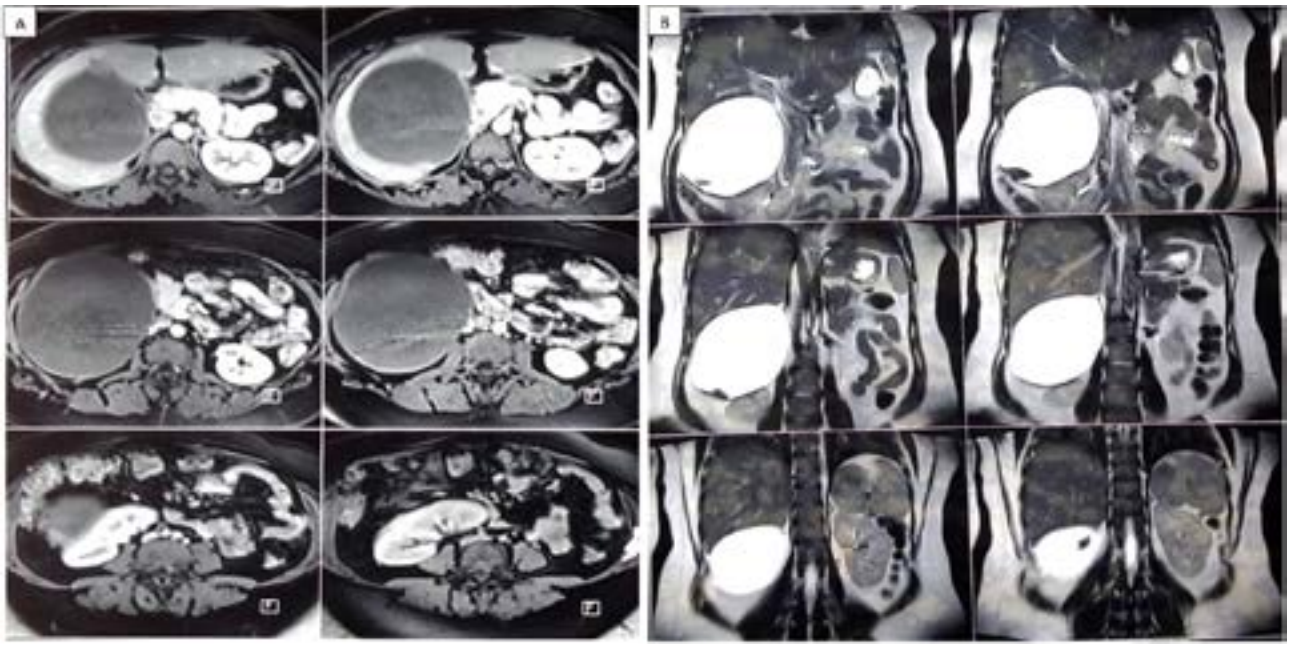

Figure 1. **T1 (A) and T2 (B) weighted images showing a thin walled retroperitoneal cystic lesion with equivocal wall enhancement, most likely to be an adrenal cyst.**

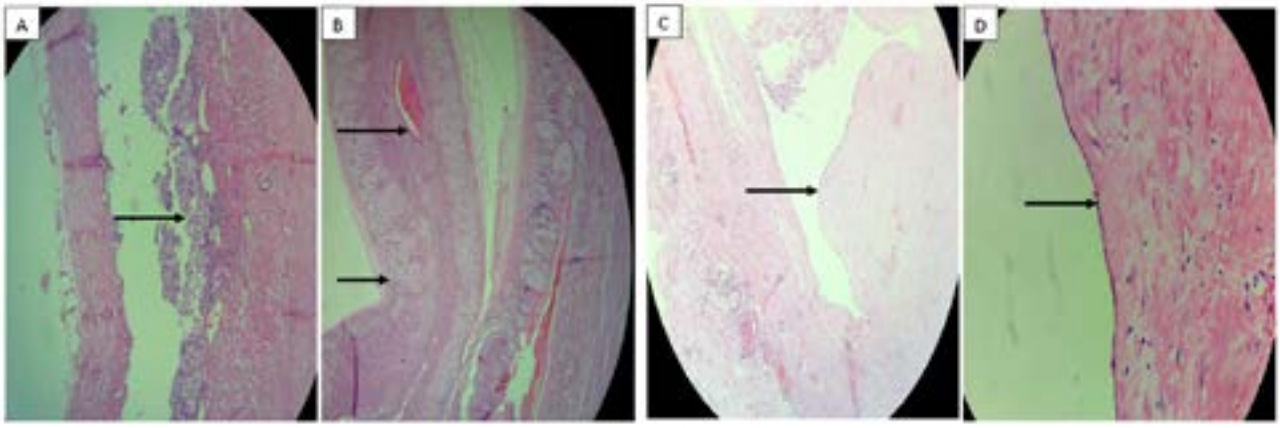

Figure 2. **A – normal adrenal gland, B – congested vessels in normal adrenal gland, C – cyst lined by flat epithelial cells, D – flat epithelial lining of the cyst (in high power)**

measuring 12 x 9.5 x 4 cms with smooth surface, shaggy inner walls and normal adrenal tissue around. The final diagnosis was adrenal cyst of epithelial type **(figure 2).**

DISCUSSION

The first case of an adrenal cyst was reported in 1670 by Greaseless.[1,2] Adrenal cysts are generally rare, 613 adrenal cysts were reported till 2004.[3] The incidence of adrenal cysts in autopsy series ranges from 0.064% to 0.18% and account for 1-22% of incidentally detected adrenal lesions.[4-6] Most of them are seen in the 3rd to 4th decades of life although cases have been reported as early as prenatal period. There is a slight female preponderance in the incidence of adrenal cysts.[7] They are highly varied in size reaching up to more than 20cms and are usually non-functional and asymptomatic. Bilateral cysts have been reported in 8-10% of cases.[8] Giant adrenal are cysts which are larger than 10cm in diameter[9] and cysts pose a diagnostic conundrum to the surgeon as localization of their origin is very difficult.

Traditionally, adrenal cysts are divided into neoplastic and non-neoplastic groups.[9] Non-neoplastic adrenal cysts may be further categorized as any of the 4 major types: endothelial or lymphangiomatous cysts (45%) pseudocyst (39%), epithelial cysts (9%) and parasitic cysts (7%) generally Echinococcal.[10-14] Endothelial cysts are the most common and include two subtypes: lymphangiomatous (42%) and hemangiomatous (3%) cysts[14] derived from their lymphatic and vascular origins respectively and are usually small measuring 0.1 to 1.5 cm in diameter.[15] Pseudocyst, the second most common pathologic type of adrenal cysts, have walls composed of fibrous tissue because they lack an epithelial or endothelial lining.[16] The aetiology of adrenal

pseudocysts is not known, however several theories have been put forward such as cystic degeneration of a primary adrenal neoplasm, vascular neoplasm and malformation as well as haemorrhage of adrenal veins into the adrenal gland.[17,18] Epithelial cysts are rare and include congenital glandular (retention) cysts, cystic adenomas, and other unspecified cysts, including mesothelial cysts.[19] Parasitic adrenal cysts may occur in association with disseminated Echinococcus infections; however, it is extremely rare for a parasitic adrenal cyst to be the only site of infection.[16,20,21]

Most adrenal cysts are asymptomatic and less than 10 cm in diameter when detected incidentally. However, symptoms occur when adrenal cysts become large enough to cause pain and gastrointestinal disturbances or when they become palpably enlarged. They may also be a consequence of intracystic bleeding or infection. Less frequent presentations include hypertension or spontaneous rupture of the cyst.[17,22]

Differential diagnosis of adrenal cysts include cystic adrenal neoplasm, lipid rich adrenal adenoma, cystic pheochromocytoma, necrotic adrenal metastasis and rare cases of subdiaphagmatic bronchogenic cyst.[23] Incidence of malignancy in adrenal cysts is reported as 7%.[24]

Routine endocrine evaluation should be performed in all cases in order not to miss a functional tumor. Owing to the rarity of occurrence of adrenal cysts, definitive diagnostic criteria based on imaging has been hard to come up with. The suggested radiographic criteria for diagnosing an adrenal cyst include a well-defined, sharply marginated mass of fluid attenuation without any evidence of enhancement. Unfortunately, radiographic criteria alone cannot rule out malignancy in adrenal cystic lesions, thus cyst aspiration or surgical excision is often performed to rule out malignancy. In small non functional tumors, aspiration of the cyst, after ruling out functional tumor, proves to be diagnostic and therapeutic at times although adequate schedule for follow up of such patients has not been defined. Though a clear aspirate usually ruled out malignancy, with the advent of radiological imaging in recent years, this practice seems outdated and obsolete.[23]

Adrenal cysts can also be incidentally detected in pregnancy.[25,26,27,28] It is usually resected electively either in second trimester or in the postpartum period as in the index case. However in rare cases it might also present as an emergency needing urgent exploration.[29] However it is worthwhile to note that the pregnancy outcome is not affected by the presence of large adrenal cysts.

Adrenalectomy is performed in most cases of large adrenal cysts to alleviate symptoms in symptomatic patients and to rule out malignancy in asymptomatic patients. Any patient with cyst size >6cm, signs of malignancy on radiological imaging, symptomatic or functional, mandates excision of the cyst. Laparoscopic adrenalectomy remains the treatment of choice in view of the advantages of the minimally invasive approach of the same, care has to be taken to abide by the oncological principles of resection, though the histopathology may turn out to be benign in most cases.

END NOTE

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Editor's Remarks: This case report is about a rare variant of adrenal cyst. The unusual fearures are that:

1. Giant cysts are very rare
2. Very few cases have been reported with presentation in pregnancy.
3. Epithelial variant is even more rare presentation of all adrenal cysts reported (9% of all adrenal cysts)

Conflict of Interest: None declared

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