ADRENAL VASCULAR (ENDOTHELIAL) CYST: A CASE REPORT AND LITERATURE REVIEW

Wattanawan EUAPITAKSAKUL[1], Surasuk TAWIPORNCHAI[2], Benjaporn CHAIWUN[3]

ABSTRACT

Adrenal vascular (endothelial) cysts are rare adrenal lesions. Herein, we reported a case of adrenal endothelial cyst in a 30-year-old pregnant woman. The patient had a 10-day history of left upper quadrant pain. Abdominal ultrasonography and computed tomography showed a huge hemorrhagic cystic lesion superior to the left kidney. Calcified cyst wall with internal septae was also noted. The microscopic examinations revealed multiloculated cysts lined focally by benign flattened endothelial cells. The general feature, differential diagnosis, pathogenesis, and management of adrenal cysts were discussed.

INTRODUCTION

Adrenal vascular (endothelial) cysts are rare.[1] They vary greatly in size from microscopic to more than 50 cm in diameter. Symptoms, particularly in large cysts, are usually associated with compression effect to surrounding visceral organs, leading to pain and vague abdominal symptoms. On the contrary, smaller ones are incidentally found during the evaluation of unrelated abdominal conditions. Clinically, the differential diagnosis of adrenal vascular cysts is diverse, including any lesion that can present with upper abdominal mass for instance, adrenal neoplasms, hepatic cysts and hemangiomas, polycystic kidneys, hydronephrosis, cystic renal cortical adenomas and carcinomas, Wilms' tumor, pancreatic, mesenteric and urachal cysts, and retroperitoneal tumors. Our aim was to report a rare case of adrenal vascular cyst recognized by ultrasonography and computed tomography.

CASE REPORT

Clinical information:

A 30-year-old woman with 15-week gestation pregnancy presented to the gynecological service with a 10 days history of left upper quadrant pain. Physical examination revealed a positive left kidney's punch. The basic laboratories were normal. The abdominal ultrasonography (US) showed a well-defined 10x15 cm thin wall cystic mass with hemorrhagic content over the left suprarenal region (figure 1). The left kidney was displaced downward to the left lower quadrant of the abdomen. Both kidneys were grossly unremarkable. The computed tomography (CT) revealed a huge 10x13x15 cm hemorrhagic cystic mass at the left suprarenal region with enhanced internal septae after contrast administration (figure 2). According to the findings, a provisional diagnosis was hemorrhagic adrenal cyst.

[1] Department of Radiology,
[2] Department of Surgery, Phayao Hospital, Meung, Phayao, 56000 Thailand
[3] Department of Pathology, Faculty of medicine, Chiang Mai University, Thailand
Running title: Adrenal endothelial cyst
Keywords: Adrenal mass, diagnosis, ultrasound, computed tomography, pathology
Corresponding author: Wattanawan Euapitaksakul, M.D. Department of Radiology, Phayao Hospital Amphur Meung, Phayao 56000 THAILAND

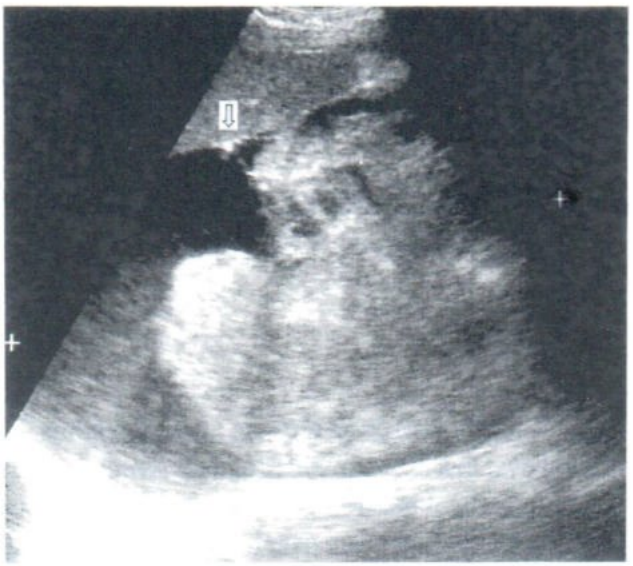

Fig. 1 Ultrasonography shows a well-defined thin wall cystic mass (arrow) with hyperechoic content. A small calcification at cyst wall is also shown.

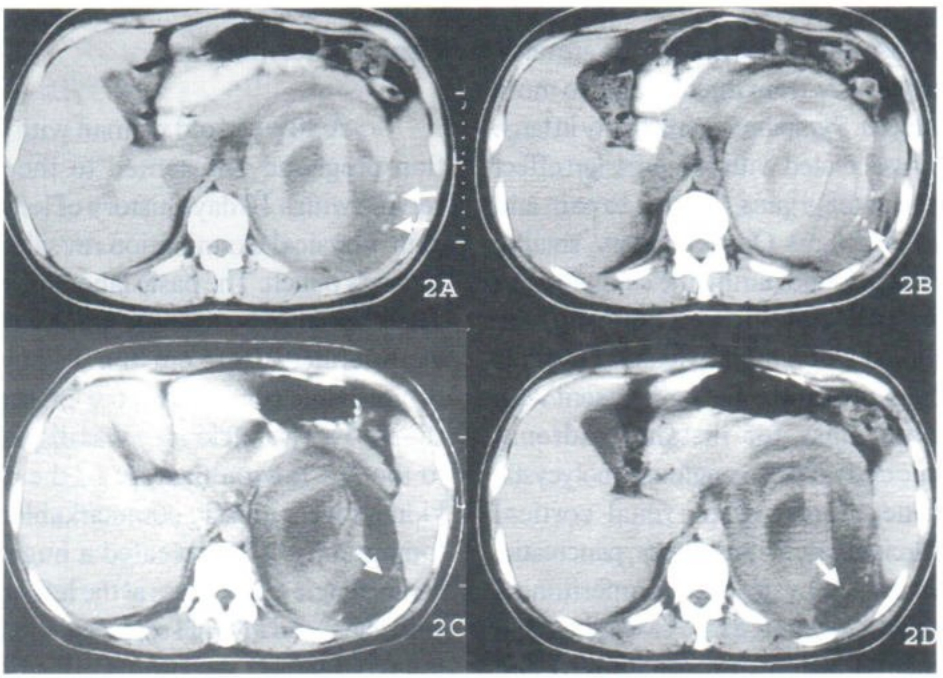

Fig. 2 (A and B) Non-enhanced CT scan through left suprarenal region shows a large heterogeneous mass at the left suprarenal region with anterior displacement of the pancreas. The high attenuation areas consistent with hemorrhage. Calcifications in the wall of the mass (arrows) are shown. (C and D) Contrast-enhanced CT scan shows a well-delineated internal septae.

Because the cyst was larger than 5 cm in diameter, an exploratory laparotomy was performed. Intraoperatively, a huge transparently thin wall cyst filled with 1300 ml of hemorrhagic fluid was found in the left retroperitoneal space. Since, the inferior margin could not be separated from the left kidney, the cyst and the left kidney were removed (figure3).

The histopathologic study revealed multiloculated cysts focally lined by benign flattened endothelial cells. The cyst contents consisted of bloody materials. The intervening stroma focally revealed benign adrenal tissue as well as focal hemorrhage. No definite malignancy was observed (figure 4).

Fig. 3 Gross pathology of adrenal cyst is shown.

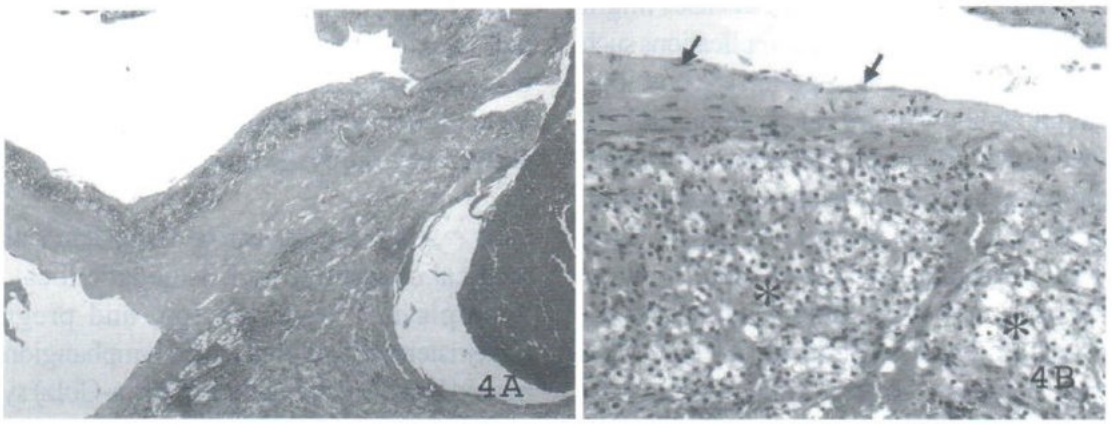

Fig. 4 Microscopic sections show multiloculated cysts (H&Ex40) (A) lined by benign endothelial cells (arrow) with sheets of benign adrenal cortical cells in the stroma (astrix) (H&Ex200) (B)

DISCUSSION

Adrenal cysts are rare tumefactive lesions, with the reported incidence of 0.06-0.18%[2] They can occur at any age, but are more commonly seen in the fifth and sixth decades of life.[1] Less than five percent of the cases occur in children.[3] Females are more affected than males. The adrenal cyst is often noted unilaterally, in which it is more commonly seen on the right side; only 5 -8% is bilateral. Symptoms and signs are usually nonspecific, such as dull flank pain, epigastric distress or indigestion, and a palpable abdominal mass.[4] Occasionally, these might accompany with constitutional symptoms, for example malaise and weakness. Fever with leukocytosis is rare. Intracystic hemorrhage can result in anemia and a slowly growing abdominal mass.[1]

Adrenal cysts have been classified into four major types, including endothelial cyst, pseudocyst, epithelial-lined or true adrenal cyst, and parasitic cysts.[1] Among those, the endothelial cysts are the most common reported (45%), in which they typically originate from lymphatic vessels, called lymphagimatous cysts (41%). However, some can rarely derived from capillaries, named angiomatous cysts. Adrenal endothelial cysts are usually incidentally found, but in exceptional cases might present with hypertension.[5] Acute complications such as hemorrhage, rupture, and infection are possible. Plain radiographs show mural calcifications in 15% of the cases.[6] Although the cyst wall is thin and smooth containing homogenous liquid contents, it, sometimes, shows slightly echoic or hyperechoic at imaging. Large cysts need to be distinguished from renal, hepatic, splenic and pancreatic cysts. Multiplanar images are helpful in localizing the origin of the cyst. The pseudocysts (39%) are the most common clinically recognized type of adrenal cyst encountered during surgery.[7] It is usually large and develops in elderly patients. However, a recent report showed giant adrenal cysts occurring in three teenage girls.[8] The walls are composed of dense fibrous connective tissue with islands of adrenal cortical tissue incorporated. Bleeding into the normal adrenal gland or tumor might be responsible for the formation of pseudocysts. This postulation is supported by a report of children having pseudocyst following adrenal hemorrhage as a result of injury or infarction in the neonatal period.[9] If the cyst is lined by epithelial, it is called true cyst, occurring in 9% of the cases. They can subdivided into true glandular or retention cysts, embryonal cysts and cystic degeneration of adrenal tumors.[10] Most of the malignant adrenal cysts are metastatic (95%), followed by pheochromocytoma (3%) and adrenocortical carcinoma (2%).[11] Malignancy should be suspected if the patients present with hormonal disturbances and hypertension. A retrospective review of the ultrasonographic feature of adrenal lesions revealed that focal absence of periglandular fat between the adrenal gland and the large vessels or liver, as well as deviation or compression of the large vessels by the adrenal lesion, may indicate malignancy.[12] Although calcification could be seen in either benign or malignant lesion, a thickened and irregular cyst wall should be suspected of malignancy. Finally, the parasitic cyst (7%) usually occurs after disseminated Echinococcus granulosus infection.[13] The content is initially purely liquid and is limited by two layers. The inner (germinal) layer is composed of the parasite, whereas the outer layer allows the passage of nutrients. The inner layer produces the laminar membrane and scoleces and secretes the cystic fluid.

Adrenal cysts can be associated with certain medical conditions such as hepatic focal nodular hyperplasia,[14] hypertension, and pregnancy.[15] Co-existence of adrenal cystic lymphangiomas with nevoid basal cell carcinoma (Gorlin-Golz) syndrome were reported.[16]

Radiological differential diagnosis of adrenal cysts should include cystic lesions in the adjacent organs such as the liver, spleen, kidneys, and cystic retroperitoneal tumors as well as other rare congeni-

tal abnormalities, such as bronchogenic cysts.[17]

Hemorrhage in adrenal gland occurs secondarily to both traumatic and nontraumatic conditions. Nontraumatic adrenal hemorrhage, however, is uncommon and may be associated with a variety of conditions including sepsis, burn, hypotension, pregnancy, cardiovascular disease, exogenous steroid, bleeding diathesis, and underlying adrenal tumors.[18] Stress events, as mentioned previously, stimulate endogenous secretion of adrenocorticotropic hormone, resulting in a marked increase in adrenal vascularity and subsequently intraglandular hemorrhage. The lesion is more commonly seen on the right side, in which this could be attributed to compression of the adrenal gland between the liver and kidney.[19] A primary adrenal cyst or tumor was reported to be the fourth most common causes of spontaneous retroperitoneal hemorrhage after renal cell carcinoma, angiomyolipoma, and renal artery aneurysm.[20] Although a cortical adenoma is the most common neoplasm of the adrenal gland, massive hemorrhage from an adenoma is extremely rare, probably due to its hypovascular nature.[21,22] Pheochromocytoma is the most common cause of massive bleeding from a primary adrenal tumor, whereas bronchogenic carcinoma is the most common cause of hemorrhagic adrenal metastases.[19]

Unfortunately, clinical and imaging findings alone and even fine-needle aspiration, cannot always make the diagnosis and differential diagnoses of these lesions. Surgical biopsy is always needed. Ultrasound, CT, and MRI have been reported having diagnostic sensitivities of 66.7%, 80%, and 100%, respectively.[23]

Management of adrenal cysts depends on clinical and imaging findings and diagnosis. Surgical excision, when possible by laparoscopic approach, is indicated in the presence of symptoms, endocrine abnormalities (even when subclinical), complications, and suspicion of malignancy. In a small cyst without evidence of malignancy, the patient may be treated conservatively with regular follow-up by ultrasonography or CT and hormonal evaluation.

In summary, a case of adrenal vascular (endothelial) cyst, a rare entity, is reported here with its radiological and pathological findings. The diagnosis of different types of adrenal cystic lesions and their differentiation from cystic lesions of adjacent organs are critical for clinical management. The combination of clinical, laboratory, and imaging findings is essential. It is important for the radiologist to be familiar with the characteristic appearance of different cystic adrenal lesions in order to guide diagnosis and patient management.

REFERENCES

1. Abeshouse GA, Goldstein RB, Abeshouse BS. Adrenal cysts: review of the literature and report of three cases. J Urol 1959;81:711-9.
2. Wahi HR. Adrenal cysts. Am J Pathol 1951 ;27: 758-61.
3. Barron SH, Emanuel B. Adrenal cyst: a case report and review of pediatric literature. J Pediatr 1961;59:592-9.
4. Travis WD, Oertel JE, Lack EE. Miscellaneous tumors and tumefactive lesions of the adrenal gland. In:Lake EE, editor. Pathology of adrenal glands. New York: Churchill Livingstone, 1990:p.351-78.
5. Gigas JM, Bucy JG, Truxler G, et al. Cystic hamartoma of the adrenal gland associated with hypertension. J Urol 1972;107:161-2.
6. Scheible W, Coel M, Siemers PT, Seigle H. Percutaneous aspiration of adrenal cysts. AJR 1977;128:1013-6.
7. Foster D. Adrenal cysts: review of literature and report of case. Arch Surg 1966; 92: 131 -43.
8. Broadley P, Daneman A, Wesson D, et. al. Large adrenal cysts in teenage girls: diagnostic and management. Pediatr Radiol. 1997; 27:550-2.

9. Zivkovic SM, Jancie-Z guricas M, Jukanovic R. Adrenal cysts in newborn J Urol 1983; 129:1031-3.
10. Kearney GP, Mahoney EM, Maher E, Harrison JH. Functioning and nonfunctioning cysts of the adrenal cortex and medulla. Am J Surg 1977;134:363-8.
11. Outwater E, Bankoff MS. Clinically significant adrenal hemorrhages secondary to metastases: computed tomographic observations. Clin Image 1989;13:195-200.
12. Besso JG, Tidwell AS, Gliatto JM. Retrospectve review of the ultrasonographic features of adrenal lesions in 21 ferrets. Vet Radiol Ultrasound 2000;41:345-52.
13. Otal P, Escourrou G, Mazerolles C, et al. Imaging features of uncommon adrenal masses with histopathologic correlation. Radiographics 1999;19:569-81.
14. Chetty R, Dada MA. Focal nodular hyperplasia of the liver coexisting with an adrenal pseudocyst. S Afr J Surg 1998;36:82-3.
15. Uretzky G, Freund H, Charuzi, luttwak EM. Cysts of the adrenal gland. Eur Urol 1978; 4:97-9.
16. Mortele KJ, Hoier MR, Mergo PJ, Ros PR. Bilateral adrenal cystic lymphangiomas in nevoid basal cell carcinoma (Gorlin-Golz) syndrome: US, CT, and MR findings. J Comput Assist Tomogr.1999;23:562-4.
17. Reichelt O, Grieser T, Wunderlich H, et al. Bronchogenic cyst: a rare differential diagnosis of retroperitoneal tumors. Urol Int. 2000; 64:216-9.
18. Rao RH, Vagnucci AH, Amico JA. Bilateral massive adrenal hemorrhage: early recognition and treatmenrt. Ann Intern Med 1989; 110:227-35.
19. Akira K, Carl MS, Randy DE, et al. Imaging of nontraumatic hemorrhagic of the adrenal gland. RadioGraphics 1999;19:949-63.
20. Swift DL, Lingeman JE, Baum WC. Spontaneous retroperitoneal hemorrhage: a diagnosis challenge. J Urol 1980;123:577-82.
21. De Resende Alves JB, de Alvarenga RJ, Kingma J, Meyer TN. Adenoma and hemorrhage of the adrenal gland. Int Surg 1976; 61:429-30.
22. Anderson WM, Timberline GA. Massive retroperitoneal hemorrhage from an asymptomatic adrenal cortical adenoma. Am Surg 1989;5:299-302.
23. Bellantone R, Ferrante A, Raffaelli M, et al. Adrenal cystic lesions: report of 12 surgically treated cases and review of literature. J Endocrinol Invest 1998;21:109-14.