Cystic Lymphangioma of the Adrenal Gland: a rare case report

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ABSTRACT

Cystic adrenal lymphangiomas are rare and benign lesions of vascular origin with lymphatic differentiation. To our knowledge, less than 60 cases are reported in the literature. We report a 37-year-old woman in whom a more than 10 cm anechoic mass with septations in right upper quadrant of the abdomen was displayed by abdominal sonogram during investigation for right flank knocking tenderness. Further computed tomography showed a large cystic mass with calcifications and enhancing septa in right adrenal gland. Subsequent operation and pathohistological examination confirmed a large cystic adrenal lymphangioma. We summarize the radiological features of the cystic adrenal lymphangiomas and differential diagnosis based on our case and the review of literature.

Lymphangiomas are rarely benign lesions of vascular origin, thought to arise from dilated lymphatic channels or developmental malformations of the lymphatic system. They mostly (95%) occur in the neck and axillary regions, rarely in the chest and abdominal cavity [1, 2]. They are extremely rare located in the adrenal gland, less than 1% of abdominal lymphangiomas originate from the adrenal gland [1]. Owing to different management and prognosis, distinguishing cystic lymphangiomas and other benign adrenal cysts from cystic adrenal malignancy on imaging are important. With the advancement of radiographic techniques, the ability to clinically characterize adrenal cystic lesions is improved. Here, we present the radiological features of cystic adrenal lymphangiomas and its common mimics to make differentiation from adrenal solid tumor with cystic change.

CASE REPORT

A 37-year-old woman presented with fever, dry cough and sore throat for three days in our emergency department. Upper respiratory infection was diagnosed. However, physical examination showed tenderness to palpation at the right flank. Initial laboratory evaluation revealed leukocytosis with elevated neutrophil count but negative finding of urinary analysis.

For evaluation of the right flank knocking tenderness, abdominal ultrasound was performed revealing a large mass measuring around 15.0 × 11.5 × 13.0 cm in size in right upper abdomen (Fig. 1). The mass are predominantly anechoic with fine septations. Subsequent abdominal CT displayed a large well-defined mass with lobulated contour in the right adrenal fossa, resulting in indentation the posterior and inferior aspect of liver surface and downward displacement of right kidney (Fig. 2). Both limbs of the right adrenal gland were elongated and wrapped the lesion. The lesion was low attenuation similar to fluid density (HU: 0-10) with mural and septal enhancement and punctate calcifications. The imaging findings favored an adrenal origin. Due to large size of the lesion and low incidence of the large benign adrenal cysts, adrenal tumor with cystic change such as cystic metastasis, cystic adrenal cortical carcinoma, pheochromocytoma with cystic change were included in the list of differential diagnosis regardless of no identifiable soft tissue component. Infectious process
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was less likely because of clear of the fat plain surrounding the lesion.

Laboratory evaluation tested for adrenal mass revealed unremarkable values for cortisol, aldosterone, ACTH, 24-h urinary excretion of vanillyl mandelic acid, epinephrine, norepinephrine, and dopamine. The functional adrenal tumor was excluded.

Surgical excision was then carried out for definitive diagnosis and also for relief of symptoms. At surgery, the large cystic mass appeared to arising from right adrenal gland which was deformed. Grossly, the cystic mass was measured around 10 × 9.8 × 7 cm (Fig. 3). The cysts had smooth and glistening inner surfaces with serous fluid or colloid content. Microscopically, sections of the lesion revealed multilocular spaces lined by flat endothelial cells which were positive for D2.40 (maker of lymphatic endothelium) and CD31 (maker of endothelial cell) (Fig. 4a, 4b, 4c). Adrenal tissue which was focally positive for synaptophysin was identified in the lesion that supported adrenal origin (Fig. 4d). Overall, the findings were consistent with cystic lymphangioma of right adrenal gland.

After a short hospital stay, the patient was discharged without complications.

DISCUSSION

Lymphangiomas are benign vascular lesions that show lymphatic differentiation. They are thought to arise from dilated lymphatic channels due to obstruction of local lymph flow or developmental malformations of the lymphatic system which failed to establish communication with the central lymphatic system [3].

They mostly (95%) occur in the neck and axillary regions; the remaining 5% are located in the lung, mediastinum, abdominal viscera mesentery and retroperitoneum [1, 2]. It is extremely rare that lymphangiomas locate in the adrenal gland, < 1% cases of abdominal lymphangiomas in one series [1]. According to the study by Carla et al. on 2011 [4], less than 50 cases are reported in the literature. To our knowledge, there are around 56 cases until 2013 based on Pubmed database. The diagnostic age ranges from 16 to 58 years old [5, 6]. The lesion size can be up to 35 cm in largest diameter [7].

Most cystic adrenal lymphangiomas are asymptomatic and discovered incidentally on radiologic studies performed for unrelated causes [4]. If symptoms do occur, they are usually related to size and position of the lesions, such as pain, gastrointestinal disturbance, or palpable mass. Acute symptoms can also occur with cystic hemorrhage, rupture, or infection [8]. Laboratory findings are nonspecific and are usually not helpful as a diagnostic tool.

Histologically, cystic lymphangiomas are thin-walled cystic masses lined with attenuated endothelial cells resembling the cells that line normal lymphatics [4]. Grossly, they present as large macroscopic interconnecting cysts, may contain chylous, serous, hemorrhagic, or mixed fluid. Histological examination and positive for CD31 and D2-40 immunostaining are diagnostic [4, 9].

The imaging features of the cystic lymphangiomas are well recognized in the neck and axillary regions. When they arise from the adrenal gland, they share similar characteristic imaging features. On sonography, cystic lymphangiomas are multilocular cystic mass or occasionally unilocular. The septations between loculi are more often thin than thick. Echogenicity depends on the nature

Figure 1

Figure 1. Sonogram of right upper quadrant shows a large multilocular cystic mass occupied right upper abdomen with close relationship with the liver. The mass is predominantly anechoic with fine septations.
of fluid are usually anechoic. If the fluid contains blood or chyle, internal echoes or fluid-debris level may be seen [10]. If mural or septal calcification is present, although uncommon, ultrasound may show acoustic shadowing [8, 11]. On computed tomography, they are characterized by lack of enhancement with intravenous contrast but may show enhancement of the cyst wall and septa [1, 8]. The fluid component is typically homogeneous with low attenuation values similar to water density, or occasionally higher if hemorrhagic or protein component is present [8].

Figure 2

Figure 2. Contrast–enhanced CT images of adrenal cystic lymphangioma with coronal view a, b, axial view c, and sagittal view d. CT images show a large well-defined cystic mass (HU: 0-10) with lobulated contours in the right adrenal fossa, resulting in indentation the posterior and inferior aspect of liver surface and downward displacement of right kidney. Both limbs of the right adrenal gland (arrow) were elongated and wrapped the lesion. Enhancement of thin wall and fine septations and punctate calcifications (arrowhead) are noted without identifiable solid component.
Negative attenuation values may occur in the presence of chyle. Calcification is identified radiographically in 15% of the cases and is typically peripheral and curvilinear [12]. Magnetic resonance imaging of simple cysts of adrenal lymphangioma can show hypointensity on T1-weighted images and hyperintensity on T2-weighted images. But more complex cysts of adrenal lymphangioma may reveal hyperintensity on both T1- and T2-weighted images [8].

The diagnostic challenge on imaging is the differentiation of cystic adrenal lymphangiomas from other benign adrenal cyst and adrenal solid tumor with cystic change including cystic metastasis, cystic adrenocortical carcinoma, and pheochromocytoma with cystic change in adult patients. Cystic lymphangiomas may be distinguished from solid tumor with cystic change by lack of enhancing soft tissue component with smooth and thin wall. The walls of solid tumor with cystic change are usually thick and irregular. Pheochromocytoma may present as cystic lesion due to central necrosis and hemorrhage. However, a near-complete cystic degeneration is rare and hormonal evaluation is useful to differentiation [13]. Differentiating cystic lymphangiomas from other benign adrenal cyst such as pseudocysts may be difficult because the imaging features of these lesions overlap [1].

Differentiating benign adrenal cystic lesions from solid tumor with cystic change is emphasized due to conservative management of the former is adequate; however, aggressive treatment of the latter is necessary. Benign features of the adrenal cystic lesion including thin wall (≤3 mm), small size (<6 cm in diameter) and homogenous near-water density can be managed conservatively and followed up with series imaging, irrespective of the presence of septa or calcifications [8, 12]. Surgery is indicated for large lesions causing symptoms, complicated cysts, functioning cysts and malignant cysts to determine diagnosis or relieve symptoms. A complete surgical excision for cystic lymphangioma is curative, although potential aggressive behavior has been described [14].

In our case, this large adrenal cystic lesion has features of cystic lymphangioma including well-demarcated border, multiloculation with thin wall and fine septations, homogeneous low density similar to water density, no identifiable soft tissue component, and uncommonly mural and septal calcification. Owing to large size of this lesion raising the possibility of malignant potential and also causing right knocking tenderness, surgical resection is performed for definite diagnosis and relief symptoms. If this lesion is smaller than 6 cm without causing symptom, conservative treatment with imaging follow up as initial management may be adequate.

In conclusion, although cystic lymphangioma is rarely arising from adrenal gland, we should particularly include cystic adrenal lymphangioma in the list of differential diagnosis for a thin-walled cystic adrenal mass with fine septations and carefully differentiate from adrenal solid tumor with cystic change.
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REFERENCES


Figure 4. Photomicrograph of histopathologic specimen. a. Low–magnification power and b. high–magnification power show multiple interconnecting cysts lined by flattened simple lining cells in submucosa (hematoxylin and eosin; ×100 and ×200). c. Positive for D2-40 immunostaining in the flattened lining cells is seen (×100). d. Focally positive for synaptophysin is identified in the lesion that supports adrenal origin (×100). Overall, the findings were consistent with benign cystic lymphangioma of right adrenal gland.