Case report

Adrenal cystic lymphangioma: A case report and review of the literature

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A B S T R A C T

A 37-year-old female was being evaluated for fever when a huge cystic lesion of about 9 × 10.5 cm² in size in the right adrenal fossa with internal septal structures containing spots of calcification was found incidentally on computed tomography. A right adrenal cyst was suspected, and right adrenalectomy and surgical resection of the lesion were then performed. The pathology showed multilocular spaces lined by flat cells, compatible with adrenal cystic lymphangioma. Immunohistochemically, the tumor was strongly positive for D2-40 and CD31. She received regular imaging follow-up, and no metastatic disease has been found until now. The radiological and pathological features of adrenal cysts and the treatment strategy for adrenal lymphangiomas are discussed.

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1. Introduction

Lymphangiomas are benign vascular lesions that are known to occur throughout the whole body. Adrenal lymphangiomas (also known as cystic adrenal lymphangiomas) are extremely rare. They were first reported in 1965, and currently less than 50 cases have been reported in the English literature.1 They are most often found incidentally as a large cystic mass, and surgical removal is necessary to rule out other types of adrenal neoplasms. Herein, we report a patient with a right adrenal cystic lymphangioma and review the radiological and pathological features of the tumor.

2. Case report

A 37-year-old Taiwanese female presented with fever off and on for 3 days. A physical examination showed RUQ (right upper quadrant) knocking tenderness, and abdominal echo revealed a huge cystic lesion with septation and a honeycomb appearance (Fig. 1). Abdominal computed tomography (CT) demonstrated a huge cystic lesion of about 9 × 10.5 cm² in size in the region between the right adrenal gland and the liver. Subtle internal septal structures with spots of calcification were seen (Fig. 2). Blood tests were all in the normal range, including a cortisol level of 10.87 μg/dL (normal range 5–25 μg/dL), ACTH (adrenocorticotropic hormone) 20.30 pg/mL (normal range 5–77 pg/mL), plasma renin activity 0.13 ng/mL/h (normal range 0.15–2.23 ng/mL/h), aldosterone 3.91 ng/dL (normal range 0.75–15.00 ng/dL), 24-hour urinary excretion of vanillylmandelic acid 2.7 mg/d (normal range 1.0–7.5 mg/d), 24-hour urinary excretion of epinephrine <3 μg/d (normal range <22.4 μg/d), 24-hour urinary excretion of normetanephrine 20.5 μg/d (normal range 11.1–85.5 μg/d), and 24-hour urinary excretion of dopamine 168.6 μg/d (normal range 50–450 μg/d).

The patient received right adrenalectomy and tumor excision in July 2011. On pathological examination, the tumor was grossly a multicystic tumor measuring 10 × 9.8 × 7 cm³ in size (Fig. 3). The inner surface was smooth and glistening, and the cysts had a serous fluid or colloid content with yellowish-orange spots in the cystic walls. There were also foci of hemorrhage and fibrosis. Microscopically, sections of the tumor revealed the features of a cystic lymphangioma composed of multilocular spaces lined by flat cells (Fig. 4). Immunohistochemically, the tumor was strongly positive for D2-40 and CD31 (Fig. 5), and the histopathological diagnosis was a cystic lymphangioma.

The patient was discharged from the hospital after 10 days without perioperative complications. Follow-up CT scans at 6 and 9 months showed a 2.9 × 2.4 cm² nonenhanced cystic lesion in the right adrenal fossa (Fig. 6). A subsequent CT scan 2 years after surgery revealed that this lesion had decreased in size to 2.2 × 1.3 cm². Postoperative seroma formation was favored, and no metastatic disease was found.
3. Discussion

Lymphangiomas are benign malformations of vessels. They are known to occur throughout the whole body, although most of the lesions are located in the neck, axilla, and mediastinum. Occasionally, they are found in the abdominal cavity (5%), including the mesentery of the small intestine, omentum, mesocolon, or retroperitoneum. The tumors can occur at all ages, and have a peak incidence from the third to sixth decades of life. The pathogenesis of adrenal lymphangiomas is still unclear; however, the most favored theory is abnormal development and/or ectasia of lymphatic vessels.

Adrenal cysts are uncommon, with a reported incidence of 0.06–0.18%. Barron and Emanuel categorized adrenal cystic lesions into pseudo-, epithelial, endothelial, and parasitic cysts. CT findings of cystic lesions vary. Pseudocysts are generally suprarenal masses with mixed density, central necrosis, or heterogeneous enhancement. Simple endothelial cysts such as lymphangiomas appear as thin-walled, fluid-density lesions on CT; they are usually hypodense and nonenhancing lesions. However, the radiological features cannot fully distinguish lymphangiomas from malignant or functional lesions such as adrenal cortical carcinoma or pheochromocytoma.

Histologically, adrenal lymphangiomas are characterized by multiloculated cystic cavities. The cysts are lined by a single layer of flat cells, and containing amorphous hypocellular fluid.

![Fig. 1. Abdominal echo: a huge cystic lesion with septation.](image1)

![Fig. 2. Computed tomography scan of the abdomen: cystic mass in the right-side retroperitoneum.](image2)

![Fig. 3. Surgical specimen: a multicystic tumor measuring 10 × 9.8 × 7 cm in size.](image3)

![Fig. 4. Sections show adrenal gland with dilated cystic spaces, lined by flat cells, and containing amorphous hypocellular fluid.](image4)

![Fig. 5. The flat lining cells are strongly positive for D2-40.](image5)
of flattened endothelial cells, and endothelial atypia or degenerative changes are uncommon. Previous reports have not shown any evidence of associated adrenal hyperplasia, adrenal adenoma, adrenal cortical adenoma, or pheochromocytoma. Immunohistochemically, cells with an endothelial lining mostly stain positively for CD31, factor VIII, CD34, and D2-40. CD31, factor VIII, and CD34 label both the vascular and the lymphatic endothelium. D2-40 is a commercially available monoclonal antibody directed against human podoplanin, a transmembrane mucoprotein that is expressed in lymphatic endothelial cells. It has emerged as an immunoreagent expressed by a variety of different neoplasms such as lymphangiomas, Kaposi sarcomas, hemangioendotheliomas, adrenal cortical carcinomas, epithelioid mesotheliomas, and seminomas. In the current case, it served as a specific marker to distinguish an angiomatous from a lymphangiomatous cyst.

Adrenal lymphangiomas are usually asymptomatic; however, nonspecific symptoms related to mass effect such as fever, abdominal pain, or palpable mass may occur. Functional lesions in endothelial cysts are not common. The management strategy is influenced by three factors: (1) functional status of the cyst; (2) the possibility of incidental malignancy; and (3) potential complications such as hemorrhage or infection. Neri and Nance reported a 7% incidence of malignancy in their review of over 600 cases of adrenal cysts from the literature. Thus, surgical removal is generally necessary to rule out other types of adrenal neoplasms. Fine-needle aspiration of fluid can be an alternative therapy if the cyst is symptomatic and nonfunctional, and raises a very low suspicion of malignancy.

Conflicts of interest

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