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CASE REPORT

Cystic Lymphangioma of the Right Adrenal Gland

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Lymphangiomas are benign malformations of the vessels. They are commonly located in the neck, axillary region and mediastinum. Lymphangioma of the adrenal gland is very rare. These lesions were first discovered as incidental autopsy findings. As the imaging techniques have improved, they now appear as incidental findings at abdominal ultrasonography and computed tomography scan examinations. They are usually asymptomatic. We present a 26-year-old woman admitted to the hospital, complaining of weakness, putting on weight, and lumbago. Her laboratory findings were within normal limits. Radiological examination revealed a 7 cm cystic lesion located in the right adrenal gland. Right adrenalectomy was performed. Histopathological examination and immunohistochemical analysis of the cystic lesion was consistent with a lymphangioma. (Pathology Oncology Research Vol 11, No 4, 242–244)

Key words: Adrenal gland, adrenal cyst, lymphangioma

Case report

A 26-year-old woman reported to the hospital, complaining of weakness, putting on weight, and lumbago. Physical examination was normal. In laboratory findings cortisol level was 1.3 μg/dl (normal range 5-25 μg/dl) and after 8 mg dexamethasone suppression test cortisol level was 2 μg/dl (normal range 3 μg/dl<), testosterone was 2.4 pg/ml (normal range 1-10 pg/ml), dehydroepiandrosteron sulfate was 80 mg/dl (normal range 35-450 mg/dl), 24-h urinary excretion of vanillyl mandelic acid was 1.6 mg/24 h (normal range 1.4-8.8 mg/24 h), 24-h urinary excretion of metanephrine was 116 μg/24 h (normal range 20-345 mg/24 h), normetanephrine was 174 μg/24 h (normal range 30-440 mg/24 h), hematocrit was 36.5%, hemoglobin was 11.1 g/dl, platelets were 229,000/μl. Abdominal ultrasonography (US) revealed a 7 cm, lobulated cystic mass with septae and sharp contours, located in the right adrenal gland. Computed tomography (CT) scan and magnetic resonance (MR) findings verified the 7 cm cystic lobulated mass associated to vena cava inferior (*Figure 1*).

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In December 2003, right adrenalectomy was performed. The patient was discharged from the hospital a week later with normal physical and laboratory findings. On pathologic examination the adrenalectomy specimen weighed 40 gram and measured 9x4x3.5 cm. On the cut surface, normal adrenal gland measured 4x0.7x0.3 cm, and a multi-septae cystic lesion of 8 cm in diameter was detected adjacent to the gland (*Figure 2*). Microscopic examination revealed a multi-cystic lesion laid with flat endothe-

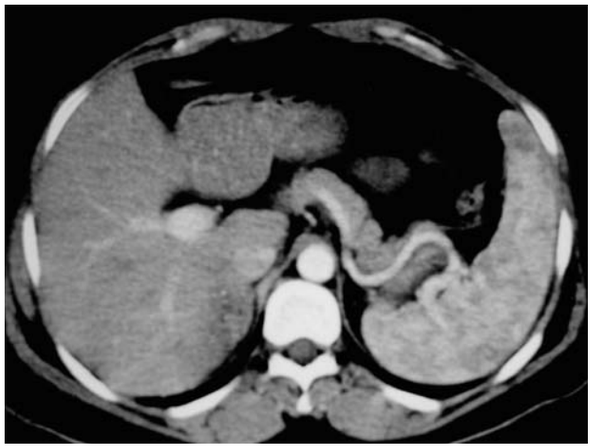

Figure 1. *Contrast-enhanced axial computed tomography image shows hypodense, lobulated solid mass of 6.5x5 cm in diameter in the right adrenal gland.*

lial cells adjacent to the normal-appearing adrenal cortex. The cystic spaces were filled with proteinous fluid (*Figure 3*). Immunohistochemically these cells stained positively for CD31 (Neomarker, clone JC/70A, 1/50 dilution) and CD34 (Neomarker, clone QBEnd/10, 1/50 dilution) (*Figure 4*). The cells were positive for smooth muscle actin (Neomarker, clone 1A4; same as asm1, 1/50 dilution), which circumscribed the cyst. Histopathological diagnosis was cystic lymphangioma in the right adrenal gland.

Discussion

Lymphangiomas are benign malformations of vessels, most frequently discovered in childhood. They are most commonly located in the neck, axillary region and mediastinum (95%). The remaining 5% are found in the abdominal cavity.[9] Four histological subtypes of lymphangiomas have been described: cystic, capillary, cavernous and vasculolymphatic malformation.[1] These types are considered as a spectrum of the same disease. Combinations of these types may be seen in the same lesion. The presence of endothelial lined lymphatic channels separated by connective tissue is the dominating histological feature.

Cysts of the adrenal glands are very rare. They were first described by Plaut, whose case studies included autopsy findings.[7,8] The incidence in autopsy studies ranges from 0.064% to 0.18%.[10,12] As the imaging techniques improved, adrenal cysts are discovered as incidental findings on abdominal US and CT examinations.[4,6,12]

Adrenal cysts have been classified into four main groups:[6] endothelial cysts (45%), pseudocysts (39%), epithelial (9%) and parasitic cysts (7%). Pseudocysts occur with hemorrhage in a normal adrenal gland or an adrenal tumor. Hemorrhage can occur secondary to trauma, bleeding disorder, burns, shock or toxemia. Epithelial cysts are comprised of cystic adenomas, glandular or retention cysts, and cystic transformation of embryonic remnants. Parasitic cysts are most commonly due to echinococcal infection with associated disseminated systemic hydatidosis. Endothelial cysts have a recognizable endothelial lining, and most of them are lymphangiomatous, however, they are not frequently encountered.[5] They are characterized by multiloculated cystic and endothelial lined cavities.[11] The endothelial lining reacts with Factor VIII-related antigen, CD31 and CD34.[2,13]

Adrenal cysts are usually asymptomatic. When they are symptomatic, symptoms are related to size and position of the cyst, and can include pain, gastrointestinal disturbance or a palpable mass. Laboratory findings are nonspecific. Rarely small adrenal cysts may be associated with Cushing syndrome, virilization or pheochromocytoma.6 The imaging methods are useful for the clinical management of an adrenal cyst. 4,10 Thin-walled (= 3 mm) cysts under 6 cm in diameter, with homogenous near-water density can be

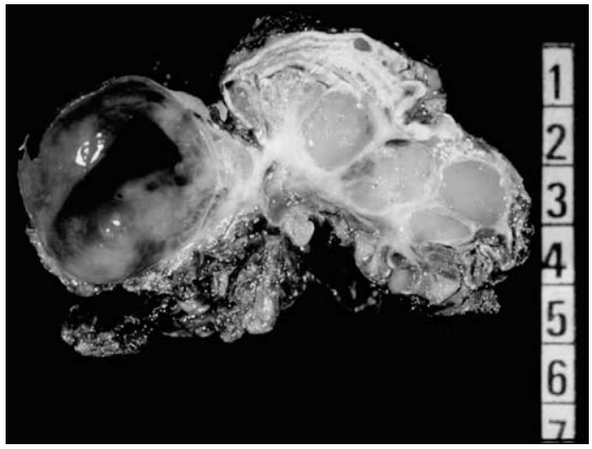

Figure 2. *Macroscopic appearance of multi-septae cystic lesion adjacent to the adrenal gland*

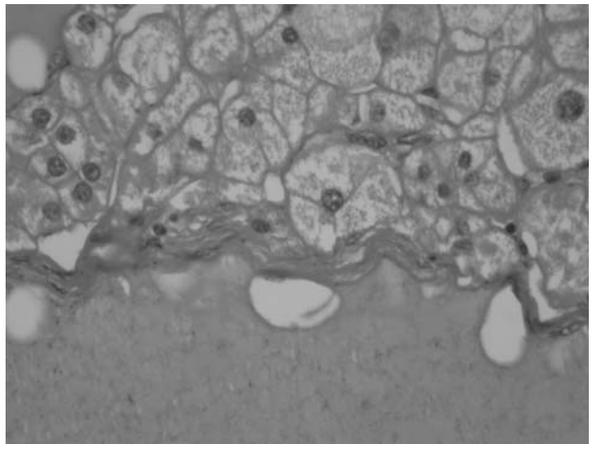

Figure 3. *Histologically cystic lesion covered by flat endothelial cells adjacent to the normal-appearing adrenal gland*

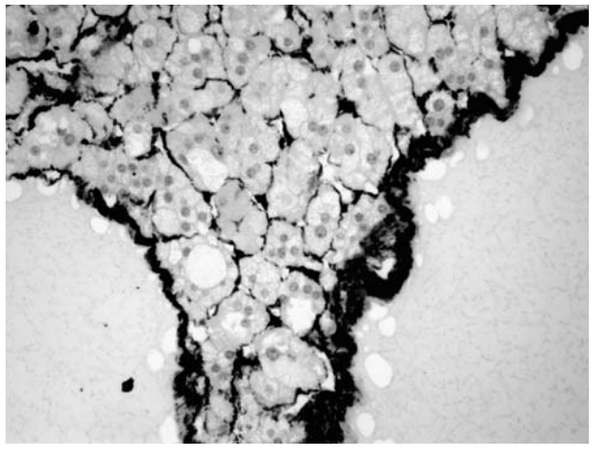

Figure 4. *Flat endothelial cells are positive for CD31.*

safely managed conservatively, irrespective of the presence of septae or calcifications.[10]

Some authors recommend aspiration of the contents of adrenal cysts for their diagnosis and management instead of surgical excision if the suspicion of malignancy is low, or the lesion is nonfunctional and asymptomatic.[3,11,14]

Potential aggressive behavior of lymphangiomas has been described, but it is very uncommon.[1,8]

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