Cystic Adrenal Lymphangioma: Differential Diagnosis

Kistik Adrenal Lenfanjiyoma

Bengü ÇOBANOĞLU, MD,[a]
Pervin KARATAŞ, MD,[a]
Selami SERHATLIOĞLU, MD,[b]
Osman DOĞRU, MD[c]

Departments of
[a]Pathology,
[b]Radiology,
[c]General Surgery,
Fırat University Faculty of Medicine,
Elazığ

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Yazışma Adresi/*Correspondence:*
Bengü ÇOBANOĞLU, MD
Fırat University Faculty of Medicine,
Department of Pathology, Elazığ,
TÜRKİYE/TURKEY
bengus_2005@yahoo.com

ABSTRACT Adrenal lymphangioma is a very rare entity. It occurs approximately in 0.06% of the general population. Because of lacking characteristic symptoms and signs and clinical insignicance preoperative diagnosis is difficult. Adrenal cysts are usually asymptomatic; if symptoms do occur, they are usually related to the mass effect and include pain, gastrointestinal disturbance, or a palpable mass. Laboratory findings are nonspecific and are usually not helpful as a diagnostic tool. A case of right adrenal cystic lymphangioma in a 68-year-old man was reported. He was admitted to the hospital with abdominal pain and abdominal distention. Ultrasound and computerized tomography (CT) scan showed the mass and suggested a diagnosis of pheochromocytoma or a metastatic mass in the right adrenal gland. The patient underwent surgery and a right adrenalectomy was performed. Histopathological examination and immunohistochemical analysis of the lesion was consistent with a lymphangioma.

Key Words: Adrenal glands; vascular neoplasms

ÖZET Adrenal lenfanjiomlar ender görülen lezyonlardır. Nüfusun ortalama %0.06'sında görülür. Preoperatif tanı, klinik seyrin sessiz olması, karakteristik bulgu ve semptomların yokluğu nedeni ile zordur. Nadiren ağrı, gastrointestinal rahatsızlık ya da kitle ile ilişkili bulgular verir. Laboratuvar bulguları nonspesifiktir ve genellikle tanı için yardımcı değildir. Bu çalışmada 68 yaşında sağ adrenal kistik lenfanjiyoma tanısı alan bir erkek hasta sunulmuştur. Hasta abdominal ağrı ve distansiyon şikâyetleri ile hastaneye başvurdu. Ultrason ve abdominal bilgisayarlı tomografide (BT) sağ adrenal bezde feokromositoma veya metastaz olabileceği öngörülen bir kitle saptandı. Hasta opere edildi ve sağ adrenalektomi uygulandı. Lezyonun histolojik ve immünohistokimyasal incelemesi lenfanjiyoma ile uyumlu bulundu.

Anahtar Kelimeler: Adrenal bez; vasküler neoplazm

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A 68-year-old male presented to the hospital with abdominal discomfort. Physical examination was insignificant. Initial laboratory evaluation revealed leukocytosis with elevated neutrophil counts. Other laboratory findings were normal. He had undergone a cholesistectomy operation 5 months ago.

A computerized tomography scan of the abdomen and pelvis showed bilateral renal cortical cysts, right adrenal mass, and nodular hyperplasia of the prostate (Figure 1). The patient was scheduled for surgery. Right adrenalectomy was performed for definitive diagnosis. Macroscopically, the ad-

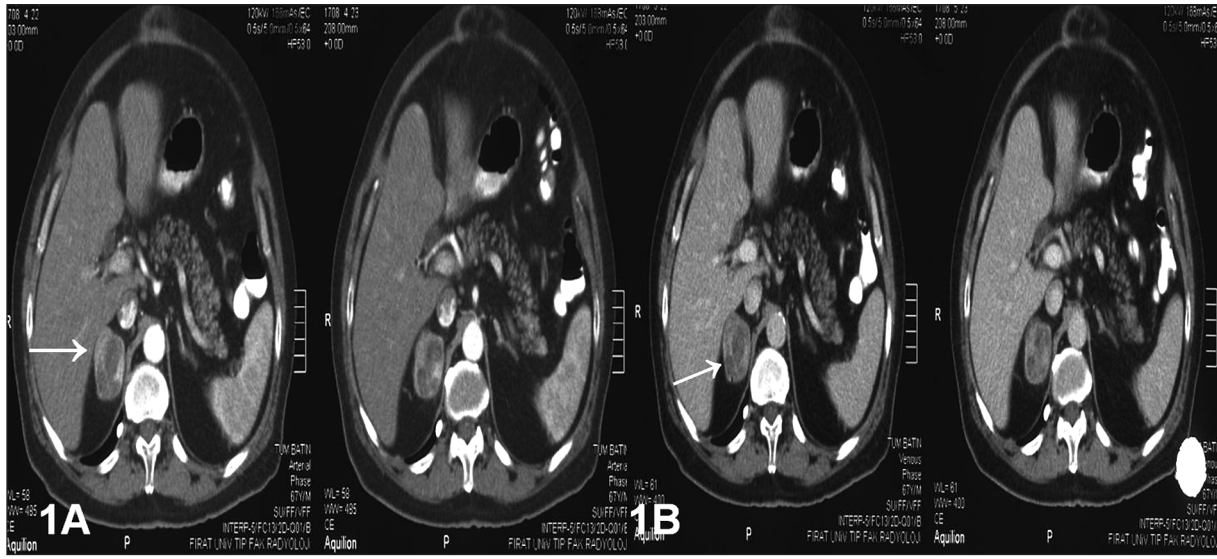

FIGURE 1: A computerized tomography scan of the right adrenal mass.

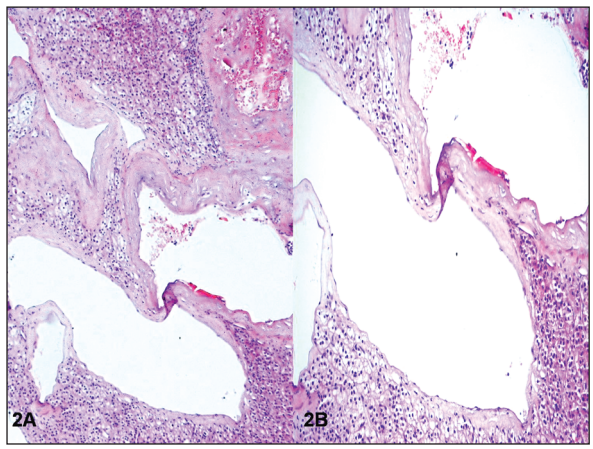

FIGURE 2: Histological examination showed irregularly shaped cystic spaces mantled by flattened endothelial cells, surrounded by normal appearing adrenal tissue (A; HE, x100, B; HE, x200).

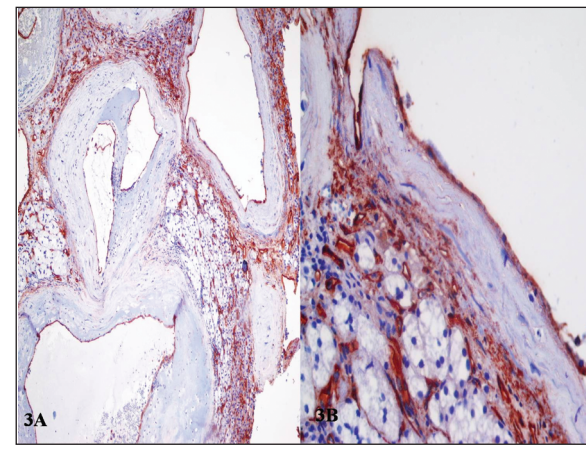

FIGURE 3: The cells lining the cyst displayed no atypia and were strongly immunopositive for CD31 (A; HE, x200, B; x400).

renal gland measured 9.5 x 5.5 x 4.0 cm with a 4.0 x 3.5 x 3.0 cm cystic component. The cut section revealed a thin walled cyst. Histological examination showed irregularly shaped cystic spaces mantled by flattened endothelial cells, surrounded by normal appearing adrenal tissue (Figure 2). The cells lining the cyst displayed no atypia and were strongly immunopositive for CD31 (Neomakers, clone JC70A) (Figure 3). Histopathologic diagnosis was cystic lymphangioma in the right adrenal gland.

Lymphangioma is a malformation of vessels and is benign. It occurs most frequently in childhood.[1] Its incidence in autopsy studies has been reported in the literature to range from 0.064% to 0.18%.[1-4] Although adrenal cysts occur at all ages, there is a peak incidence between the third to sixth decades.[2,5] Lymphangiomas are most commonly located in the neck, axillary region and mediastinum (95%). The remaining 5% are found in the abdominal cavity.[1] They are typically unilateral with equal distribution on each adrenal. The female to male ratio is 2:1.[2] Adrenal cysts are usually asymptomatic. When they are symptomatic, symptoms are related to the size and position of the cyst. Laboratory findings are nonspecific. Rarely, adrenal

cysts may be associated with Cushing syndrome, virilization or pheochromocytoma.[1]

Four histological subtypes of lymphangiomas were described; cystic, capillary, cavernous and vasculolymphatic malformation.[1,2] Adrenal cysts were classified into four main groups: endothelial cysts (45%), pseudocysts (39%), epithelial cysts (9%), parasitic cysts (7%).[5]

Endothelial cysts include angiomatous, lymphangiomatous, and hamartomatous cysts. Lymphangiomatous cysts are characterized by multilocutated cystic and endothelium lined cavities. The endothelial lining reacts with Factor VIII related antigen, CD31 and CD34.[1] Pseudocysts occur with hemorrhage into a normal adrenal gland or adrenal tumor. Epithelial cysts are comprised of cystic adenomas, glandular or retention cysts, and cystic transformation of embryonal remnants. Parasitic cysts are most commonly due to echinoccocal infection.[2]

Clinical management of an adrenal cyst can be planned in the view of imaging findings. If a small and asymptomatic lesion is identified in the adrenal gland, it can usually be followed up with serial imaging.[5] Management of larger or symptomatic lesions may require surgical resection.

On CT, uncomplicated adrenal cysts are characterized by lack of enhancement with intravenous contrast.

On magnetic resonance imaging, adrenal cysts are low in signal intensity on T1 weighted images and high on T2 weighted images.

On ultrasound, the diagnosis of cystic lymphangioma is suggested by the presence of a well marginated, anechoic lesion, located at the suprarenal area.[1,2]

Although lymphangiomas are benign lesions, very uncommonly aggressive behavior potential in lymphangiomas also has been described.[1]

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