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Case Report

Ectopic adrenocortical adenoma in the renal hilum mimicking a renal cell carcinoma ☆

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ABSTRACT

Ectopic adrenocortical tissue can arise along the path of embryonic migration, such as the celiac axis, broad ligament, adnexa of the testis, and spermatic cord. Occasionally, ectopic adrenocortical tissues undergo marked hyperplasia and develop into ectopic adrenocortical adenomas. This report describes the case of a 60-year-old man who was incidentally found to have a lipid-containing mass with early enhancement and delayed washout in the right renal hilum. A renal cell carcinoma was suspected, and robot-assisted partial nephrectomy was performed, but the final diagnosis was an ectopic adrenocortical adenoma. We should include ectopic adrenocortical adenoma in the differential diagnosis when we find a lipid-containing tumor adjacent to the kidney.

Introduction

Ectopic adrenocortical tissue is a developmental abnormality of the adrenal gland. It can arise along the path of embryonic migration, such as the celiac axis, broad ligament, adnexa of the testis, and spermatic cord [1]. Occasionally, ectopic adrenocortical tissues undergo marked hyperplasia and develop into ectopic adrenocortical adenomas and carcinomas [2]. A case of an ectopic adrenocortical adenoma in the renal hilum that mimicked a renal cell carcinoma (RCC) is reported.

Case report

A 60-year-old man presented to our hospital for further examination of a nodular shadow in the left middle lung field found on a chest X-ray performed during a general checkup. He had a history of hypertension diagnosed at the age of 55 years and had received drug therapy from a local physician. His blood pressure was 137/80 mm Hg, and the remainder of the physical examination was unremarkable. Serum sodium and potassium levels were normal.

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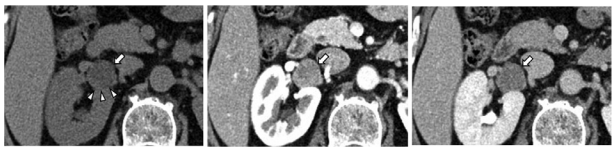

Fig. 1 – CT of the abdomen. Noncontrast CT image (A) shows a low-density mass in the right renal hilum (arrow). The mass is partially in contact with the right renal cortex (arrowheads). Dynamic contrast-enhanced CT demonstrates slight enhancement in the arterial phase (B) and washout in the delayed phase (C).

We re-examined the chest X-ray, but no nodular or mass shadow was found. Noncontrast chest computed tomography (CT) also showed no remarkable findings in his lungs. However, a well-defined, homogeneous, low-density mass (CT value: 15 Hounsfield Units), 2.3 × 2.3 × 2.1 cm in size, was incidentally found in the right renal hilum. The mass was partially in contact with the right renal cortex. Dynamic contrast-enhanced abdominal CT demonstrated that the mass showed slight enhancement in the arterial phase and washout in the delayed phase (Fig. 1). Bilateral adrenal glands showed no abnormalities. There was no continuity between the adrenal gland and the mass. On magnetic resonance imaging (MRI), T1-weighted gradient echo images showed a uniform decrease in signal intensity on the out-of-phase images compared with the in-phase images, which indicated distribution of a microscopic fat component within the mass (Fig. 2). On the apparent diffusion coefficient maps derived from diffusion-weighted imaging (DWI) with a b value of 1000 s/mm^2, the mass showed lower intensity than the renal cortex. Based on these imaging findings, RCC was diagnosed because the mass was thought to originate from the renal cortex, and have microscopic fat component.

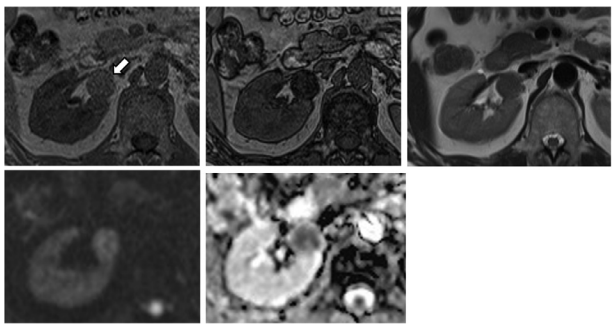

Fig. 2 – MR imaging of the abdomen. Axial T1-weighted gradient echo in-phase image (A) shows slight high intensity in the mass (arrow), and the out-of-phase image (B) shows decreased signal intensity compared with the in-phase image. On T2-weighted imaging (C), the mass shows slight hypointensity, and the capsular structure is unclear. Diffusion-weighted imaging (DWI) with a b-value of 1000 s/mm^2 (D) shows slightly higher intensity, and the apparent diffusion coefficient (ADC) maps derived from DWI (E) show lower intensity than the renal cortex.

Robot-assisted partial nephrectomy was performed. Intraoperative findings showed that the mass was in contact with the right kidney, as suggested by the imaging findings. However, the mass was loosely attached to the kidney, and briefly removed. Although there was no apparent residual lesion on the surface of the kidney, a part of the kidney in contact with the mass was resected. The histopathological findings showed an adrenocortical neoplasm without capsule, and the tumor was distant from the native adrenal gland. According to the Weiss criteria, the adrenocortical neoplasm was benign, and the Ki-67 labeling index was approximately 1%. The final diagnosis was an ectopic adrenocortical adenoma arising from adrenal rest tissue in the renal hilum (Fig. 3).

The patient's postoperative course was uneventful. Blood pressure and serum electrolyte levels did not change significantly. There was no recurrence at evaluation 2 years after surgery.

Discussion

A case of an ectopic adrenocortical adenoma in the renal hilum that mimicked an RCC was described. To the best of our knowledge, there have been few cases of ectopic adrenocortical adenomas located in the renal hilum reported in the literature.

The adrenal gland consists of 2 parts, the cortex, and medulla. The adrenal cortex arises from the coelomic mesoderm of the urogenital ridge at the fifth week of gestation. By the eighth week of gestation, the cortical mass separates from the rest of the mesothelial tissue. The adrenal medulla arises from neural crest tissue in the adjacent sympathetic ganglion at the level of the coeliac plexus. The neural crest tissues migrate toward the adrenal cortex at the seventh week of gestation, and they gradually invade the cortex.

Ectopic adrenal glands may form when fragments of the cortical cells break off during development. The ectopic adrenal gland may contain cortex and medulla or only cortex, depending on whether the fragments break off before or after the invasion of the medullary tissue into the cortex. Ectopic adrenal tissue is found in up to 50% of neonates, but most of them regress in early infancy [3]. The prevalence of ectopic adrenal tissue in adults was reported to be 1% [4]. The ectopic adrenal tissue can occur anywhere in the migration path descending with the gonads, including in the retroperitoneum, testis, broad ligament, kidneys, ovaries, and inguinal region. The most common reported sites of ectopic adrenal tissue are the celiac axis region (32%), broad ligament (23%), adnexa of the testis (7.5%), and the spermatic cord (1.2%) [1]. An ectopic adrenal gland has been rarely reported in the lungs, stomach, and even in the intradural space and brain [5].

Occasionally, ectopic adrenocortical tissues undergo marked hyperplasia and develop into ectopic adrenocortical adenomas and carcinomas [2]. Although most ectopic adrenal tissues were non–functional, few ectopic adrenocortical tumors can produce hormones such as cortisol, aldosterone, or androgens, which can lead to the patient presenting with Cushing's syndrome or hyperaldosteronism, or it can produce extreme virilization [6]. The patient in the present case had

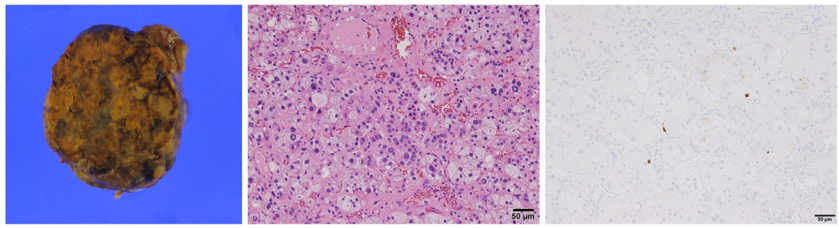

Fig. 3 – Yellowish-brown 2.3 × 2.3 × 2.0 cm solid tumor (A). Microscopically, the tumor is composed of clear cells and compact cells, which is consistent with adrenal gland origin. (hematoxylin-eosin staining; magnification: 200 × [B]). The Ki-67 labeling index is approximately 1% (magnification: 200 × [C]) (Color version of the figure is available online.)

hypertension, but corticosteroid levels were not measured because an ectopic adrenocortical tumor was not included in the differential diagnosis before surgery.

Diagnostic imaging, such as CT and MRI, is important to detect ectopic adrenocortical tumors. Although it is challenging to diagnose an ectopic adrenocortical adenoma preoperatively, imaging features of adrenocortical adenoma could assist in the diagnosis. On noncontrast CT, an adrenocortical adenoma often appears low density due to its high lipid content [7]. A dynamic study often shows rapid contrast enhancement and washout [8]. On MRI, chemical shift imaging is a reliable sequence for diagnosing adrenocortical adenoma, especially when it is not hypodense on noncontrast CT. Chemical shift images show decreased signal intensity on the out-of-phase image compared with that of the in-phase image, which indicates a microscopic fat component within the adenoma [9]. However, these imaging features also apply to RCC. In previous studies, a signal intensity decrease was observed in 60%-80% of clear cell RCC due to its intracellular lipid[10,11]. Other than clear cell RCC, microscopic fat is also found in fat-poor angiomyolipoma (AML), papillary RCC, chromophobe RCC, and oncocytoma [11]. Macroscopic fat is found in classic AML, Wilms tumor [12], and rarely in RCC [13]. On DWI, the ADC values are reported to not be valuable in the differentiation of adrenocortical adenomas, and metastases because of the overlapping of ADC values [14].

To date, there have been 6 cases of ectopic adrenocortical adenoma in the renal hilum [6,15–19]. The patients (2 males, 4 females) ranged in age from 27 to 63 years, with tumor sizes of 2.5-5.3 cm. Of the 6 cases, 3 had continuity with the kidney [6,15,16], and 5 reported endocrine abnormalities, including Cushing's syndrome [6,15–17], borderline elevation of testosterone [18], and hyperaldosteronism [6]. Most of them were correctly diagnosed due to their endocrine abnormalities. The remaining one case was a non–functioning ectopic adrenocortical adenoma in the left renal hilum [19]. The possibility of ectopic adrenocortical adenoma was not considered due to the lack of hormonal abnormalities. Because the mass was clearly located apart from the renal parenchyma, a benign retroperitoneal tumor was suspected. In the present case, the mass was thought to originate from renal cortex, and there were no symptoms to suspect hormonal abnormalities other than hypertension. Therefore, it was difficult to differentiate an ectopic adrenocortical adenoma from clear cell RCC preoperatively.

In conclusion, a case of an ectopic adrenocortical adenoma in the renal hilum that mimicked an RCC was described. Although less frequent, we should include ectopic adrenocortical adenoma in the differential diagnosis when we find a lipid-containing tumor adjacent to the kidney. Furthermore, measurement of hormone levels could assist in making the diagnosis.

Patient consent

Informed consent was obtained from the patient for the publication of this case report.

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