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Case Report

Concurrent renomedullary interstitial cell tumor and angiomyolipoma: a case report

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Abstract: Renomedullary interstitial cell tumor (RMICT) is a commonly incidentally found benign tumor at autopsy. However, RMICT is rarely recognized in routine surgical pathological specimens. Renal angiomyolipoma (AML) is also an uncommon entity of renal parenchymal tumor. To the best of our knowledge, there were only three reported cases with synchronous RMICT and AML in the English literatures. Herein, we described a patient with concurrent RMICT and AML in the same kidney. This is a 36-year-old asymptomatic woman presented to our hospital due to incidentally found right renal tumor by routine health checkup. During the surgery for the main renal tumor, a tiny RMICT (0.4 cm in diameter) was also identified by the surgeon.

Keywords: Angiomyolipoma, kidney, renomedullary interstitial cell tumor

Introduction

Renomedullary interstitial cell tumor (RMICT) is a benign tumor of renal parenchyma, which is localized in the medulla [1]. RMICTs are commonly found incidentally at autopsy [2]. However, it is rarely identified as a renal mass clinically, therefore, rarely is resected. Angiomyolipoma (AML) is also a relatively uncommon benign mesenchymal tumor of renal parenchymal neoplasm, which is characterized by co-expression of smooth muscle and melanocytic markers [3]. Synchronous RMICT and AML are even much rarer. To the best of our knowledge, only three cases were reported in the English literatures to date [4, 5]. Herein, we described an extremely rare case with coincidence of renal AML and RMICT.

Case report

A renal tumor approximately 3 cm in size was incidentally found by routine health checkup in a 36-year-old woman. The patient had neither systemic disease nor family history of tuberous sclerosis. Computed tomography (CT) scan of abdomen discovered a 3.3 cm renal tumor at lower pole of right kidney (**Figure 1A**). The impression of CT scan was renal cell carcinoma with preliminary staging T3aN0M0. Therefore, partial nephrectomy was performed for treatment. During the operation, a tiny nodule, which was not visible in the CT scan, was also incidentally identified. Both the main tumor and tiny nodule were totally resected, and the specimens were sent for pathological examination.

Materials and methods

The specimens were fixed in 10% formalin solution and embedded in the paraffin block. Sections were cut and stained with hematoxylin and eosin for light microscopy. Immunohistochemical (IHC) stains were performed using standard reagents and techniques on a BOND-MAX Automated Staining System (Leica Microsystems, Wetzlar, Germany). Briefly, sections were deparafinized, hydrated, and subjected to heat-induced antigen retrieval with Bond Epitope Retrieval Solution (EDTA based pH 9.0 solution, Leica Microsystems). The primary antibodies, including smooth muscle actin (clone αsm-1, Novocastra™, Newcastle Upon Tyne, United Kingdom, 1:200), S-100 protein (clone S1/61/69, Novocastra™, 1:500), HMB45 (clone HMB45, Novocastra™, 1:100), and Melan A (clone A103, NovocastraTM, 1:100) were applied for thirty minutes at room temper-

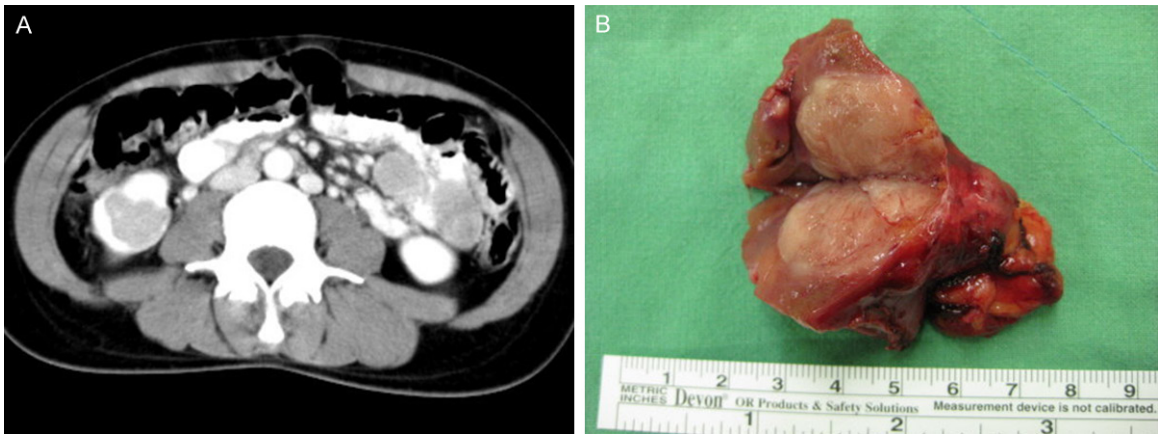

Figure 1. A. Computed tomography scan of abdomen revealed a 3.3 cm renal tumor at the right kidney. B. On sectioning, a well-circumscribed, solid and tan renal tumor was identified.

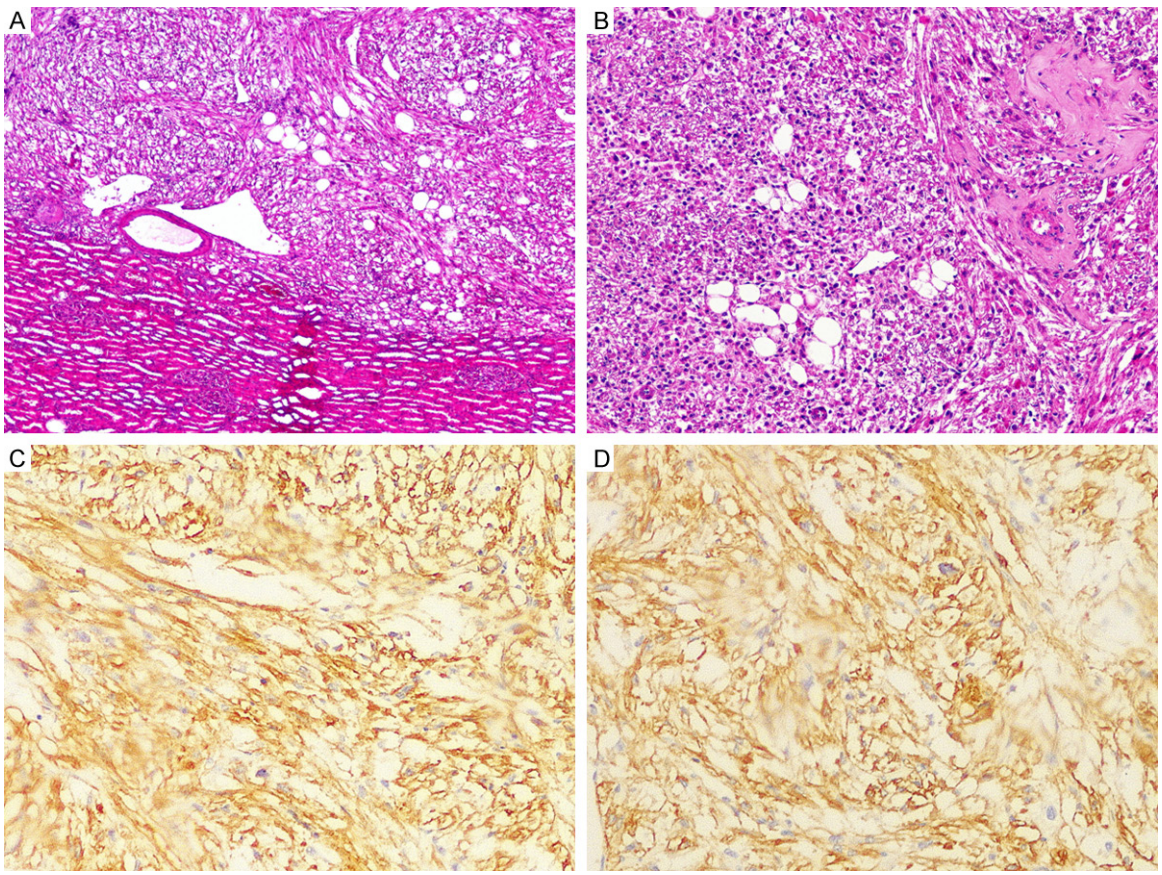

Figure 2. Histopathology and immunohistochemical profiles of the angiomyolipoma. (A) The tumor was well demarcated and non-encapsulated. (B) It composed of three components, including mature adipose tissue, thick-walled blood vessels and spindle to epithelioid smooth muscle-like cells. Note the radial pattern of the smooth muscle-like cells emanating from the blood vessel walls. The tumor cells were strongly immunoreactive to (C) smooth muscle actin and (D) HMB45. (A: ×40, B: ×100, C and D: ×200).

ature followed by application of biotin-free Bond Polymer Refine Detection Novocastra (Leica Microsystems). Appropriate positive controls for each antibody and negative control

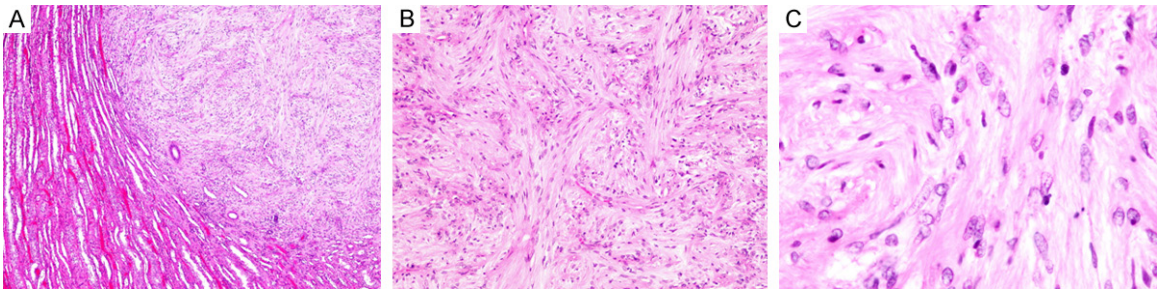

Figure 3. Histopathologic characteristics of the renomedullary interstitial cell tumor. A. The tumor was well demarcated and located at the medulla of kidney. A few renal tubules were entrapped at the periphery. B and C. The stellate to polygonal tumor cells exhibited bland round to ovoid nuclei in a loose faintly basophilic myxoid stroma. (A: ×40, B: ×100, C: ×400).

were run in parallel as manufacture's instruction.

Results

On bisection of the kidney, a well-demarcated and tan solid tumor measuring 3.5 × 3.2 × 3.0 cm was found (**Figure 1B**). The isolated tiny nodule was submitted separately and it measured 0.4 × 0.3 × 0.3 cm. Microscopically, the main tumor was well demarcated and non-encapsulated and showed classic triphasic histology: mature fat, thick-walled blood vessels and spindle to epithelioid smooth muscle-like cells (**Figure 2A**, **2B**). By immunohistochemistry, the tumor cells were immunoreactive to both smooth muscle markers (smooth muscle actin) and melanocytic markers (HMB45 and Melan A), consistent with an angiomyolipoma (**Figure 2C**, **2D**). The tumor cells were immunostained negative for S100 protein. The isolated tiny nodule was located at the renal medullary pyramid (**Figure 3A**). It was also well circumscribed and composed of small stellate or polygonal cells in a background of loose faintly basophilic myxoid stroma (**Figure 3B**, **3C**). A few renal medullary tubules were entrapped in the matrix at the periphery (**Figure 3A**). During the regular follow-up of eight months, neither recurrent nor metastatic disease was identified.

Discussion

Renomedullary interstitial cell tumor (RMICT) was firstly described by Lerman et al. in 1972 [2]. Originally, it was nominated as fibroma of renal medulla. RMICT is the most common renal tumor in adulthood. They are usually identified incidentally in autopsy. The prevalence of RMICT in autopsy series ranges from 16% to 41.8% and it is more prevalent in the elderly [6]. However, it rarely draws clinicians' attention because of usually an asymptomatic tumor and too small size to identify by imaging. Therefore, RMICTs are uncommonly recognized clinically and then excised. Although almost all RMICTs are tiny nodules within a renal medullary pyramid, extremely rarely, "giant" tumor large enough to be observed clinically and resected surgically was also ever reported [7]. The cell origin of RMICT is normal medullary interstitial cells, which regulate water and salt absorption, blood flow, and eventually blood pressure by secretion of prostaglandin, synthesized by cyclooxygenase-2 (COX-2) [8]. In the study of Gatalica et al., concomitant expression of COX-2, microsomal prostaglandin E synthase-1, and prostaglandin E2 receptor were observed in most RMICTs, implicating the possible autocrine growth loop in the pathogenesis of RMICTs [9].

Renal angiomyolioma (AML) is also an uncommon renal neoplasm, which account for about 1% of surgically resected renal tumors [3]. AML is a benign mesenchymal tumor with classical "triphasic" pattern, composed of adipose tissue, spindle to epithelioid smooth muscle cells and thick-walled blood vessels. AML features co-expression of both smooth-muscle and melanocytic markers, and therefore is included in the family of neoplasms with perivascular epithelioid-cell differentiation (PEComas) [10].

The coincidence of RMICT and AML is even rarer. To the best of our knowledge, only three cases in the English literatures were reported [4, 5]. In the report by Mustafa-Guguli et al. [5], there were even four synchronous tumors in bilateral kidney. Clear cell renal cell carcinoma

along with small angiomyolipoma and renomedullary interstitial cell tumor were found in the right kidney, while chromophobe renal cell carcinoma arose from the left kidney.

In summary, we have reported a rare case of concurrent angiomyolipoma and incidentally found renomedullary interstitial cell tumor in the same kidney. Both tumors are uncommon renal mesenchymal tumors in surgically resected specimen and exhibited typical histopathologic features, cytomorphology and immunohistochemical stains. To the best of our knowledge, the present case is the fourth case with synchronous renomedullary interstitial cell tumor and angiomyolipoma.

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Disclosure of conflict of interest

None.

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