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Simultaneous occurrence of renal cell carcinoma and angiomyolipoma in the same kidney

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ABSTRACT

Introduction: Renal cell carcinoma and angiomyolipoma are well-known tumors of kidney. However, coexistence of these tumors in the same patient is a rare condition. Synchronous occurrence of renal cell carcinoma and angiomyolipoma is more commonly seen in patients with tuberous sclerosis. Case Report: A 76-years-old female was admitted to urology clinic with a complaint of flank pain. She had pain and tenderness on her right flank on physical examination. Radiological imaging revealed a solid mass in right kidney. She underwent right radical nephrectomy. Conclusion: We report a case of clear cell renal cell carcinoma and angiomyolipoma occurring in the same kidney without stigmata of tuberous sclerosis and with the review of related literature.

Keywords: Angiomyolipoma, Renal cell carcinoma, Simultaneous neoplasms

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INTRODUCTION

Renal cell carcinoma (RCC) is a common neoplasm in general population. Clear cell (conventional) type, the most common, accounts for approximately 60% of all renal tumors. It affects primarily adults and shows male predominance. Angiomyolipoma is an uncommon benign mesenchymal tumor of kidney. Sporadic form is nearly 1% of surgically removed renal tumors but it may be seen more commonly in association with tuberous sclerosis (TS). It preferably affects female adults in sporadic form, but there is no sex predilection in TS patients [1]. However, coexistence of these tumors was reported in limited number in literature. We represent such a unique case in this report.

CASE REPORT

A 76-year-old female was admitted to urology clinic with a new onset of right flank pain. On physical examination, she had tenderness and pain on her right flank area. Laboratory results showed minimal inflammation and blood urea nitrogen level was elevated. Abdominal CT revealed a solid mass in lower pole of the

right kidney with cystic necrotic areas showing irregular inferior margin with an exophytic extension. Besides, there were many cysts located in the parapelvic region on radiological examination. The patient underwent to right radical nephrectomy with suspect of renal tumor.

Surgical specimen weighted 485 g and was 20x9x6.5 cm in size with perirenal adipose tissue. On cross section, two well-defined lesions were seen in the lower pole of the kidney (Figure 1). Main lesion had solid cystic appearance and dirty yellow color and it was 5 cm in diameter. It had a small protrusion in the inferior part but it did not exceed to the renal capsule. The other lesion, misconceived as an extension on imaging, was 3 cm in diameter with solid and yellowish appearance and separated from main lesion with a definite margin. Even though radiological imaging did not disclose any difference between these two lesions, each of them has distinct histological features on microscopical examination. Main lesion consisted of tumor cells in solid sheets, in which occasional dilated acinar structures formed a cystic pattern. The tumor cells had round, uniform nuclei with finely granular open chromatin and inconspicuous nucleoli and mainly clear cytoplasm surrounded by definite margins. Besides, regular small thin walled vessels were observed throughout the lesion (Figure 2A). It was diagnosed with conventional type RCC (Fuhrmann grade 2). This lesion was stained positively with CD10 and Vimentin by immunohistochemistry. The other lesion was separated by a thin fibrous capsule from the main lesion (Figure 2B) It was composed of mature adipocytes, smooth muscle cells and poorly organized thick walled blood vessels (Figure 2C). It was diagnosed with angiomyolipoma. Immunohistochemically, angiomyolipoma was stained by HMB-45, while there was no expression of HMB45 in RCC. Surgical margins including ureter and renal vein were free of tumor. The patient had neither lymph node nor distant organ metastasis.

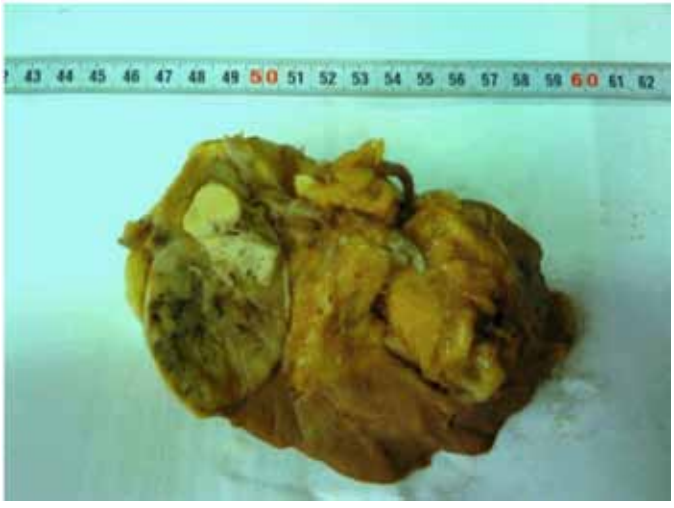

Figure 1: Macroscopic appearance of surgical specimen.

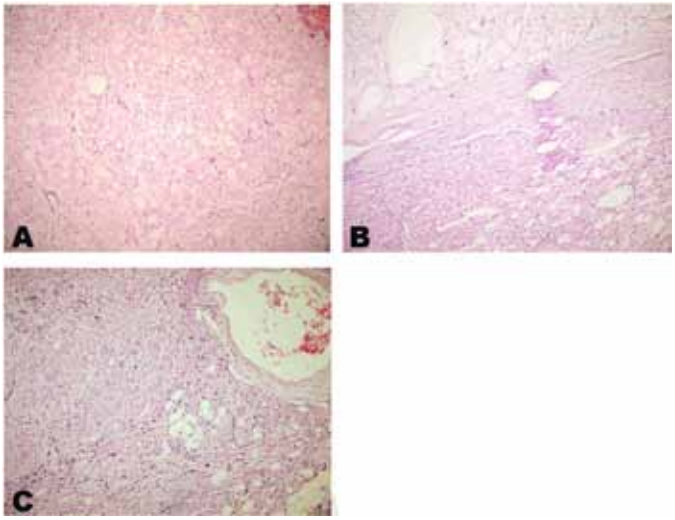

Figure 2: Microscopic appearance of collision tumor. (H&E: Hematoxylin & Eosine staining). (A) Renal cell carcinoma: malignant tumor composed of cells with oval nuclei, inconspicuous nucleoli and clear cytoplasm, (H&E stain, x200). (B) Note the thin fibrous capsule separating two neoplasms. (H&E stain, x100), and (C) Angiomyolipoma: benign tumor composed of a mixture of adipocytes, blood vessels and smooth muscle (H&E stain, x200).

DISCUSSION

Conventional type RCC is the most common and poorly understood tumor in both clinical and pathological aspects. Hereditary factors may play an important role in development of RCC and also RCC can be associated with familial disorders. In our case, there was no known family history of renal tumor or related familial diseases and hereditary disorders. Like RCC, angiomyolipoma can be associated with TS, an autosomal dominant disorder. Although angiomyolipoma shows higher incidence in association with TS, both RCC and angiomyolipoma can be associated with TS. The synchronous presentation of renal cell carcinoma and angiomyolipoma was described in the literature in both healthy patients and patients with TS [2–23]. In 2001, Jimenez et al. denoted that approximately 50 cases had been reported in the literature [2]. They also reported coexistent RCC and angiomyolipoma of 23 cases in a study of 36 cases, which is also summarized in Table 1 [2]. In addition to that, we listed 24 more cases, including our case in Table 1 [3–23].

Although RCC affects males twice as frequently as females in sporadic cases, it is seen more commonly in female patients with TS [2]. However angiomyolipoma has no female predominance in TS patient. Our patient was a woman but had no clinical signs of TS. Jimenez et al. suggested that coexistent neoplasms were more frequently in woman. In concurrent tumors, angiomyolipoma is mostly found incidentally [2]. The data in literature indicate that there is no considerable difference in coincidence of these tumors between in patients with and without TS (14 patients with TS, 31 patients without TS: see Table 1). Besides, left kidney is likely to be involved. Even though it was not indicated

Table 1: Documented cases of coexistence of renal cell carcinoma and angiomyolipoma in the same kidney.

Year	Author	TS*	Site of tumor	Tumors	Number of the cases
2000	İzaki et al. [4].	+	Not available	AML* + granular RCC*	1
2000	Bragagnolo et al. [5].	-	R*	AML + clear cell RCC + cortical adenoma	1
2001	Val-Bernal et al. [6].	-	L*	AML+ clear cell RCC	1
2001	Jimenez [2]	+	Not stated separately, but mentioned left predominancy	AML+ clear cell RCC	6
		-		AML + clear cell RCC	9
		-		AML + papillary RCC	2
		-		AML + chromophobe RCC	4
		-		AML + collecting duct RCC	1
		+		AML+ unclassified RCC	1
2002	Mai et al. [7].	-	Not stated	AML+ papillary (chromophil) RCC	1
		-	Not stated	AML + RCC	1
2003	Jun et al. [8].	-	L	AML + clear cell RCC+chromophobe RCC	1
2004	Billings et al. [9].	-	Not available	AML+ RCC	1
2005	Kida et al. [10].	+	R	AML + granular cell RCC	1
2006	Inci et al. [11].	-	L	AML + adenocarcinoma	1
2006	Rotman et al. [12].	-	Allograft kidney	AML + papillary RCC (type 1)	1
2006	Morelli et al. [13].	-	L	AML + Oncocytoma + chromophobe RCC	1
2007	Inomoto et al. [14].	-	R	AML + clear cell RCC	1
2007	Corsenca et al. [15].	+	L	AML+unclassified RCC	1
2009	Capaccio et al. [3].	Not stated	Not stated	AML + clear cell RCC	1
2009	Radopoulos et al. [16].	-	R	AML + Oncocytoma + chromophobe RCC	1
2009	Khallouk et al. [17].	+	R	AML+clear cell RCC	1
2010	Aron et al. [18].	Not stated	Not stated	AML+clear cell RCC	2
2010	Kang et al. [19].	+	L	AML+ chromophobe RCC+ clear cell RCC	1
2012	Cao et al. [20].	-	L	EpithelioidAML+clear cell RCC	1
2013	Konosu-Fukaya et al. [21].	-	L	AML+papillary RCC	1
2013	Behnes CL et al. [22].	+	L	AML+unclassified RCC	1
2013	Hussain et al. [23].	+	L	AML+ clear cell RCC	1
2014	Kokenek-Unal et al.	-	R	AML + clear cell RCC	1

*TS: Tuberous sclerosis, AML: Angiomyolipoma, RCC: Renal cell carcinoma, R: Right kidney, L: Left kidney

separately in all reports, the most common subtype of RCC, coexists with angiomyolipoma was clear cell RCC, with 26 cases. This may be easily explained by the fact that clear cell RCC is the most common subtype. Since the management of treatment differs in different renal tumors, interpretation of radiological imaging takes a critical place in surgical approach [3]. A huge angiomyolipoma may mislead to diagnosis of a RCC, or in the same manner a blood vessel rich RCC may imitate an angiomyolipoma. In our case, preoperative imaging did not rule out the presence of different lesions and angiomyolipoma was found incidentally. Although it is a rare occasion, the coexistence of RCC or even malignant transformation should be considered in follow-up of angiomyolipoma patients, particularly with TS.

CONCLUSION

In summary, we represent an uncommon case of concomitant renal cell carcinoma and angiomyolipoma in the same kidney. The presence of any pathogenetic relationship between these two tumors still remains unclear. We listed a series of cases and hope that our report will be helpful for other researchers in further investigation of simultaneous renal neoplasms.

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Author Contributions
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Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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