Unilateral simultaneous renal oncocytoma and angiomyolipoma: case report

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Abstract

A rare case of synchronous angiomyolipoma and oncocytoma in the same kidney of a 70 year old man is presented. A left renal mass was found incidentally by ultrasound. Computerized tomography and magnetic resonance imaging revealed a 1.3 cm mass in the mid-portion of the left kidney, whereas on the lower pole of the same kidney, a 3.3 cm mass was also revealed, consistent with angiomyolipoma. A working diagnosis of renal cell carcinoma was made. A radical nephrectomy was performed. Microscopically, the tumor of the lower pole was found to be an angiomyolipoma, whereas the mid-portion tumor was an oncocytoma. Until now, only 16 cases of unilateral simultaneous presence of renal angiomyolipoma and oncocytoma have been reported. Of these cases, all except one were female and three were associated with the tuberous sclerosis complex. It is well worth remarking, that renal oncocytoma overlap with other renal neoplasms, therefore nephrectomy remains the treatment of choice.

Renal angiomyolipoma and oncocytoma are uncommon neoplasms and their simultaneous presence in the same kidney is rare. Only 16 cases have been reported until now in the literature. The purpose of this paper is to present an additional case without evidence of tuberous sclerosis.

Introduction

Renal angiomyolipoma and oncocytoma represent uncommon neoplasms and their simultaneous presence in the same kidney is rather rare. To the best of our knowledge, only 16 cases have been reported in the literature [1-3]. Oncocytoma, originating from renal tubular cells, is a relatively recently reported benign epithelial tumor that accounts for about 5% of surgically resected renal neoplasms in adults [4]. The nomenclature of angiomyolipoma was first introduced by Morgan et al in 1951[5] to describe a renal tumor that contained a berrant vasculature with variable amounts of intermixed smooth muscle and adipose tissue. Renal angiomyolipoma, generally of embryonal cell origin, represents less than 1% of all surgically removed tumors and is frequently associated with tuberous sclerosis [6]. Oncocytomas have also been associated with cortical adenomas and renal cell carcinoma, whereas angiomyolipomas have been associated with...
renal cell carcinomas, a papillary adenoma, a cystic nephroma and a metanephric adenoma [1,7,8].

We present a case report of renal angiomyolipoma and oncocytoma without evidence of tuberous sclerosis.

**Case presentation**

**Clinical Case**
The patient was a 70 year old asymptomatic male with a history of cholelithiasis in whom a solid 1,3 cm medial left renal mass was diagnosed incidentally by ultrasound. There was no significant past medical history, specifically seizures or mental retardation. On physical examination, head, neck, heart and lungs were normal. Neurological testing gave normal results. Blood renal tests were normal. Computed tomography (CT) showed a 1,3 cm well-defined, exophytic solid mass without cystic characteristics in the mid-portion of the left kidney; whereas on the lower pole of the same kidney, a 3,3 cm mass that contained a significant amount of fat was revealed. This mass was radiographically compatible to an angiomyolipoma. Invasion or infiltration into the perinephric fat, collecting system of vessels or regional lymphadenopathy and metastases were not encountered. On subsequent magnetic resonance imaging (MRI), the two renal masses were confirmed. In the mid-portion of the left kidney a 1,3 cm well-defined, homogenous mass was described, which appeared hypointense relatively to the renal cortex on T1-weighted images and isointense on T2-weighted images (Figure 1). No central scar was detected. Furthermore, the presence of an angiomyolipoma in the left lower lobe was confirmed (Figure 2). The right kidney was unremarkable radiographically. A working diagnosis of renal cell carcinoma was made. The patient underwent a left radical nephrectomy through left subcostal incision and recovered uneventfully.

**Pathologic Findings**

**Macroscopic examination**
On gross examination, at the lower pole of the kidney an ovoid, yellowish 2,8 × 1,8 × 1,3 cm lesion which seemed to invade in the perinephric fat was found. In addition, there was a firm, well-encapsulated, brownish 1,7 × 1,2 × 1,0 cm tumor in the mid-portion of the kidney. Careful sectioning of the kidney did not reveal any additional lesion.

**Microscopic examination**
In the mid-portion of the kidney, the neoplasm exhibited a uniform population of plump cells arranged in alveolar-type nests and trabeculae with a granular, acidophilic cytoplasm. The morphological features were those of an oncocytoma (Figure 3). The lower pole tumor had features of an angiomyolipoma with predominant lipomatous and myomatous components. Thick-walled vessels and areas of hemorrhage within the tumor were present. Small foci of epithelioid cells without nuclear pleomorphism or increased mitotic activity were also noted (Figure 4).

Figure 1
Magnetic resonance imaging: Well-defined, homogenous mass in the mid-portion of the left kidney.

Figure 2
Magnetic resonance imaging: An angiomyolipoma of the left lower lobe.
Immunohistochemistry carried out on the sections of the oncocytoma showed negativity for antibody for vimentin and a weak positivity for antibody for CK 7.

The final confirmed diagnoses included an angiomyolipoma of the lower pole and oncocytoma of the mid-part of the left kidney.

**Discussion**

Renal oncocytomas, first described by Zippel in 1942 [9], are benign, relatively non-frequent neoplasms, and they represent 3.2 to 5% of all primary renal masses arising from intercalating cells of the cortical collecting ducts. Oncocytoma is considered to be a benign neoplasm in the majority of cases; this is the reason why there is only one documented case of liver metastasis in literature [10]. However, it has also been reported that oncocytomas may occasionally involve fat tissue in up to 20% of cases and lymphovascular structures in up to 5% [11]. Most of these tumors are single and unilateral, although bilateral and multifocal cases have also been described [10]. Cytogenetically, there is not much evidence in the available literature, but it is possible to subdivide oncocytomas into three families, as far as their genetic abnormalities are concerned: oncocytomas with losses of chromosomes 1 or Y; oncocytomas with balanced translocations involving 11q13 (region encoding mitochondrial DNA) and oncocytomas with miscellaneous abnormalities [12]. Clinically, oncocytomas may be asymptomatic, but symptomatic patients may present with initial signs of haematuria, flank pain or palpable mass. The diagnosis of these benign lesions is generally achieved by computed tomography (CT) or magnetic resonance imaging (MRI). The appearance of a typical central stellate scar can occasionally be mimicked by necrosis in a renal cancer and this feature is not considered specific [13]. The characteristic histological features of oncocytomas are the dense eosinophilic cytoplasm which may be exclusively or predominantly granular, generally uniform nuclei, and abundant mitochondria [10]. Considering the lack of diagnostic yield and low sensitivity of biotical procedures and imaging techniques, it is well worth remarking that renal oncocytoma may overlap with other renal neoplasms, such as renal clear cell carcinoma with oncocytic features [14]. Therefore, surgical excision/nephrectomy remains the treatment of choice [15].

Renal angiomyolipomas are uncommon, benign tumor-like formations, consisting of mature adipose tissue, smooth muscle cells and thick-walled blood vessels and account for less than 1% of surgically removed tumors. The perivascular epithelioid cell is thought to be the cell of origin of angiomyolipomas [16].

A common immunophenotype of these tumors is the consistent expression of melanoma-associated antigens, particularly HMB-45. Angiomyolipomas are commonly associated with tuberous sclerosis, which is an autosomal dominant disorder characterized by seizures, mental retardation, skin lesions and hamartomatous lesions in many organs [17]. Rarely, angiomyolipomas can be associated with von-Hippel Lindau disease, von Recklinghausen syndrome, and autosomal dominant polycystic kidney disease [18]. However, about 80% of renal angiomyolipomas occur sporadically and develop predominantly in women between the fourth and seventh decade.
of life, and are usually large, symptomatic, single and uni-
lateral. In contrast, angiomyolipomas associated with
tuberous sclerosis most often occur at a younger age and
they tend to be asymptomatic, small, multifocal and bilat-
eral [19]. Our patient, had no obvious clinical manifesta-
tions of tuberous sclerosis. Multiple renal angiomyolipomas, according to the criteria set forth by
Gomez [20] are considered diagnostic of tuberous sclero-
sis complex. Mett产学 evaluation of the nephrectomy
specimen in our case, however, did not reveal additional
foci of angiomyolipoma. The epithelium lining the cysts
in tuberous sclerosis has been described as morphologi-
cally distinctive; that is, the lining cells are large and often
strongly eosinophilic with large, hyperchromatic nuclei
and usually identifiable mitoses [17]. Nevertheless, these
features were not present in microscopic sections in our
case. Therefore, we regarded the angiomyolipoma in our
case as sporadic (non- tuberous sclerosis associated).

The diagnosis or renal angiomyolipoma is usually straight
forward due to its characteristic sonographic appearance
and its typical CT morphology. Ultrasonographically,
most angiomyolipomas are hyperechogenic lesions
resembling the echogenicity of the renal sinus fat; the fat
component accounts for the negative attenuation values
on CT scans and the hyperintensity on T1- weighted MR
images [21]. However, these tumors can mimic a renal cell
carcinoma, particularly if the fat content is low or if the fat
is obscured by blood.

As the risk of haemorrhage in angiomyolipomas less than
4 cm in diameter is minor, the surgical treatment in
asymptomatic cases is generally reserved for those tumors
that are greater than 4 cm. Surgical excision, including
radical nephrectomy, may remain the treatment of choice;
nonetheless, in the appropriate setting, partial nephrec-
tomy, enucleation, or wedge resection can be alternative
options [21].

Although concurrence of renal cell carcinoma and onco-
cytoma within the same kidney is well recognized [19],
the simultaneous unilateral existence of angiomyolipoma
and oncocytoma is uncommon. To our knowledge, only
16 cases [1-3] of concurrent angiomyolipoma and onco-
cytoma, including 3 cases in association with tuberous
sclerosis, have been reported in the literature, some as iso-
lated case reports and others as collective series encom-
passing other tumors [1]. In cases where the relevant
information was available, the age of the patients ranged
from 42 to 73 years. Most cases occurred in women with
only one case occurring in a man. This observation paral-
lels the female predominance of angiomyolipoma as
opposed to the male predominance of oncocytoma. Our
patient was 70 years old, which fits in with this profile, but
was a male, which makes our case remarkable.

The size of both tumors varied greatly and in some
patients the presence of two tumors was not detected pre-
operatively [2]. In patients where the preoperative radiog-
raphy was described, the presence of renal cell carcinoma
could not be excluded. In our case, the angiomyolipoma
measured 2, 8 cm and the oncocytoma was 1,7 cm. Both
of them were revealed on abdominal imaging, but the
radiologic evaluation could only clarify the type of the
angiomyolipoma due to its characteristic radiologic fea-
tures. However, the other lesion was felt to be a renal cell
carcinoma. Therefore, a radical nephrectomy was indi-
cated in our patient.

Conclusion
In conclusion, we present an additional case of an onco-
cytoma associated with an angiomyolipoma, which is
remarkable due to the fact that it was detected in a male
patient. With respect to preoperative diagnosis, these neo-
plasms remain a challenge for the surgeon because they
mimic renal cell carcinoma in the majority of cases. Our
case did not occur in the setting of tuberous sclerosis. Of
the 16 cases reported in the literature, only three cases
associated with tuberous sclerosis complex have been
documented. However, if angiomyolipomas are found
incidentally in nephrectomy specimens along with other
tumors, tuberous sclerosis should be excluded retrospec-
tively.

Consent
Written informed consent was obtained from the patient
for publication of this case report and accompanying
images. A copy of the written consent is available for
review by the journal’s Editor-in Chief.

Competing interests
The authors declare that they have no competing interests.

Authors’ contributions
TT - responsible for critical revision of scientific content;
AY - drafted the manuscript; MK - contributed substan-
tially to manuscript conception and design; GA - assisted
in the preparation of the manuscript; EKV - performed
histopathological and immunohistochemical analyses
and contributed to the pathology content; ND - have
made substantial contributions to manuscript conception
and acquisition of data; IV - the surgeon, approved the
final version of the manuscript for publication; All
authors read and approved the final version of the manu-
script.

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