SPORADIC RENAL ANGIOMYOLIPOMA WITH
CONCURRENT ANEURYSM FORMATION

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Angiomyolipoma is the most common mesenchymal renal neoplasm; it is observed in 0.1 to 0.22% of the general population. The classic form of angiomyolipoma has triphasic histopathologic characteristics, containing variable proportions of dysmorphic blood vessels, smooth muscle, and adipose tissue. Angiomyolipomas that usually are found in patients with tuberous sclerosis tend to bleed because of their hyper-vascularity and the presence of small aneurysms. CT is the diagnostic tool of choice because of its high sensitivity for detecting fat; thin section non-contrast scans are preferable for measurement of fat density. We present the case of a 35-year-old female patient referred to the emergency room for colicky pain, with occasional irradiation to the right upper quadrant. An abdominal ultrasound was ordered under the suspicion of cholelithiasis; it revealed a mass in the upper lobe of the right kidney. A supplementary abdominal MSCT obtained in both nephrographic (80-120 secs) and excretory (3-5 mins) phase reported a hypodense mass located at the level of the upper pole of the right kidney. A renal angiogram with embolisation of the tumour and aneurysm was performed, and then the patient underwent right open radical nephrectomy. In this report, we discuss the imaging features of the angiomyolipoma as well as the two-step treatment using transarterial embolisation of the tumour and aneurysm followed by radical nephrectomy.

Keywords: angiomyolipoma, aneurysm, embolisation, kidney neoplasm, nephrectomy.

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СПОРАДИЧЕСКАЯ ПОЧЕЧНАЯ АНГИОМИОЛИПОМА
С ВНУТРИПУХОЛЕВОЙ АНЕВРИЗМОЙ

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Aнгиомиолипома является наиболее распространенным мезенхимальным новообразованием почек и наблюдается в 0,1%-0,22% от общей численности населения. Классическая форма ангиомиолипомы имеет трехфазные гистопатологические характеристики, содержащие различные пропорции дисморфных кровеносных сосудов, гладких мышц и жировой ткани. Ангиомиолипомы которые обычно встречаются у пациентов с туберозным склерозом, часто кровоточат из-за гиперваскуляризации и наличия небольших аневризм. Компьютерная томография является «золотым» стандартом в диагностике данной патологии в виду высокой чувствительности метода.

Представляем клинический случай 35-летней пациентки, которая обратилась в

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отделение неотложной помощи по поводу болей в эпигастральной области. УЗИ брюшной полости было проведено при подозрении на желчнокаменную болезнь, при этом в верхней доле правой почки было выявлено образование. По данным МСКТ брюшной полости в нефрографическую (80-120 сек) и в экскреторную (3-5 мин) фазы выявлено гиподенсное образование, расположенное на уровне верхнего полюса правой почки. Была выполнена селективная ангиография с эмболизацией опухоли и аневризмы с последующей правосторонней радикальной нефрэктомией. В этом наблюдении представлены особенности визуализации ангиомиолипом, а также возможности лечения с использованием трансартериальной эмболизации опухоли и аневризмы с последующей радикальной нефрэктомией.

Ключевые слова: ангиомиолипома, аневризма, эмболизация, новообразования почек, нефрэктомия.

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Introduction.

Angiomyolipoma is the most common mesenchymal renal neoplasm; it is observed in 0.1 to 0.22% of the general population and is four times more common in female [1]. The classic angiomyolipoma has triphasic histopathologic characteristics, containing variable proportions of dysmorphic blood vessels, smooth muscle, and adipose tissue [2]. Two types have been described: sporadic angiomyolipoma and angiomyolipoma associated with tuberous sclerosis, these types reveal a distinctly different pathology and imaging appearance, and in some cases, a distinct clinical behaviour which determines the treatment (Table 1) [3]. Angiomyolipomas in patients with tuberous sclerosis tend to bleed because of their hypervascularity and the presence of small aneurysms. Macroaneurysms, are rarely seen in imaging of these patients [4].

Angiomyolipomas may present as a well-defined focal and infiltrative mass or as a pedunculated mass [5]. Angiomyolipomas, more than 4 cm in diameter, require treatment because of the risk of bleeding [6]. CT is the diagnostic tool of choice because of its high sensitivity detecting fat; thin section non-contrast scans are preferable for measurement of fat density [6]. AML appears hyperechoic on sonography equal to renal sinus fat or producing negatively attenuating areas on CT or T1 hyperintensity with signal loss on fat-saturated sequences on MR imaging [7, 8]. With the intent of early curative surgery for a presumed renal cell carcinoma, many renal masses are resected at surgery without a confirmed diagnosis. Many of them are benign, and some are angio-

myolipomas. The determination of renal angiomyolipoma using imaging is, therefore, as important as ever [3]. Surgery is the best treatment for these cases because of the risk of severe bleeding following aneurysmal rupture [5]. This report describes an unusual clinic presentation of renal angiomyolipoma with an aneurysm found in a 3D CT reconstruction in a 35-year-old female.

Case report.

A 35-year-old female was referred from the emergency room with colicky pain, predominantly in the epigastrium, intermittent, without triggers, with occasional irradiation to the right upper quadrant and sometimes bilateral lumbar pain, which decreased partially with the use of oral analgesics. Later, the patient presented an episode of severe pain accompanied by diaphoresis and fainting, reaching syncope.

The patient referred non-quantified weight loss in the last three months and progressive fatigue. Blood studies were unremarkable, hematuria was observed in the urinalysis, with preserved renal function. An abdominal ultrasound was ordered, and it revealed a mass in the upper lobe of the right kidney. A supplementary abdominal CT obtained in both nephrographic (80-120 secs) and excretory (3-5 mins) phase reported: right kidney in a usual situation with dimensions 11.3x4.8x4.8 cm; a hypodense mass was observed in the parenchyma with a hypodense centre located at the level of the upper pole of right kidney, measuring 8.4x6.1x7.0 cm. In the non-contrasted phase, mass density was between -71 to 23 Hounsfield

Table №1. Classification and clinical features of renal angiomyolipoma.

		Classification of the renal angiomyolipoma	imaging features of renal angiomyolipoma types		Pathological behaviour	Management
			CT	MRI		
Sporadic (80%)	Triphasic benign AML	Classic AML	Fat attenuation	Signal loss on FS MRI and CS MRI	Benign	Observation
		Fat Poor AML	No evidence of fat at unenhanced CT**			
		Hyperattenuating AML	Hyperattenuating (> 445 HU) homogeneously enhancing	T2-hypointense No signal loss on CS MRI No signal loss on FS MRI	Benign	Biopsy followed by observation
		Isoattenuating AML	Isoattenuating (- 10HU-45 HU)	T2-hypointense Signal loss on CS MRI + /- Signal loss on FS MRI***	Benign	Biopsy followed by observation
	AML with epithelial cysts		Hyperattenuating (445 HU), homogeneously enhancing with cystic areas or multilocular cystic	T2-hypointense, homogeneously enhancing with cystic areas, or multilocular cystic	Benign	Biopsy followed by observation
	Epithelioid AML		Hyperattenuating, (445 HU) heterogeneously enhancing or multilocular cystic	T2-hypointense heterogeneously enhancing or multilocular cystic	Potentially malignant	Resection or mTOR inhibitor
Syndromic (20%)	AML in tuberous sclerosis complex				Benign and potentially malignant	Relative to sporadic types, more likely to need treatment including resection, embolisation, ablation or mTOR inhibitor
	AML in lymphangioleiomyomatosis				Benign and potentially malignant	Similar to sporadic types, but mTOR inhibitor can be one choice

AML: angiomyolipomas; FS: frequency-selective fat suppression; CS: chemical shift; mTOR (mammalian target of rapamycin). Attenuation values are approximations.

When thin (1.5–3 mm) sections are used; *Signal loss on FS MRI may or may not be present.

Fig. 1. MSCT. A 35 years old female, renal angiomyolipoma.

A, B, C - axial CT views during the non-contrast, nephrographic and excretory phases respectively, revealing a heterogeneous, predominantly hypodense mass located at the level of the upper pole of right kidney.

D, E - coronal and sagittal views with contrast enhancement depicting the renal mass in the upper pole of the right kidney.

F - 3D reconstruction where it is possible to identify the presence of a renal aneurysm (white arrow).

Рис. 1. МСКТ. Женщина 35 лет, почечная ангиомиолипома.

А, Б, В – аксиальные реконструкции, нативная, нефрографическая и экскреторная фазы. Определяется гетерогенное, преимущественно гиподенсное образование, расположенное на уровне верхнего полюса правой почки.

Г, Д - корональная и сагиттальная реконструкции, с контрастным усилением. Определяется образование верхнего полюса почки.

Е - 3D реконструкция. Аневризма почки (белая стрелка).

Fig. 2 a (Рис. 2 а)

Fig. 2 b (Рис. 2 б)

Fig. 2 c (Рис. 2 в)

Fig. 2 d (Рис. 2 г)

Fig. 2 e (Рис. 2 д)

Fig. 2 f (Рис. 2 е)

Fig. 2 g (Рис. 2 ж)

Fig. 2 h (Рис. 2 з)

Fig. 2 i (Рис. 2 и)

Fig. 2. Angiography (A-F), gross specimen (G-I). Renal angiomyolipoma, A 35 years old female.

A-F - selective injection into the right-renal artery to visualise de angiomyolipoma and aneurysm.

A, B, C - show the gradual filling of the vascularity of the right kidney and the aneurysm inside the tumour; the mass is located in the upper pole of the kidney (black arrow) there are small intratumoral vessels (white arrowheads).

D, E, F - images after endovascular treatment demonstrating the absence of filling in the aneurysm and tumoral vessels with preservation of the middle and lower poles of the kidney; black arrowheads point the vascular contour of the tumoral mass.

G-I - excised right kidney.

G - external view after the radical nephrectomy showing preservation of the renal capsule.

H, I - internal view after sagittal cut of the kidney revealing the tumoral mass with the presence of a non-ruptured aneurysm (white arrow), it is possible to see the limits of the tumoral mass and the normal parenchyma with sparing of the inferior vena cava (black arrow).

Рис. 2. Ангиограмма (А-Е), макропрепарат (Ж-И). Ангиомиолипома почки, женщина 35 лет.

А-Е - селективная ангиография правой почечной артерии для визуализации ангиомиолипомы и аневризмы.

А, Б, В - постепенное заполнение сосудистой системы правой почки и аневризмы внутри опухоли; образование расположено в верхнем полюсе почки (черная стрелка), имеются небольшие внутритухольные сосуды (белые стрелки).

Г, Д, Е - изображения после эндоваскулярного лечения, демонстрирующие отсутствие заполнения аневризмы и опухолевых сосудов с сохранением среднего и нижнего полюсов почки; черные стрелки указывают на сосудистый контур опухолевой массы.

Ж-И – макропрепарат правой почки.

Ж - состояние после радикальной нефрэктомии, показывающий сохранение почечной капсулы.

З, И - внутренний вид после сагиттального разреза почки, выявляющий опухолевую массу с наличием неразрывавшейся аневризмы (белая стрелка), визуализируются границы опухолевой массы и нормальной паренхимы почки с сохранением нижней части полой вены (черная стрелка).

units; it was associated with a heterogeneous pattern of infiltration fat. In the retrospective evaluation of the 3D reconstruction of the CT, an image suggestive of the aneurysm was observed (Fig. 1A-F). A selective right-renal angiogram with embolisation of the tumour and aneurysm was performed (Fig. 2A-F). Based on the clinical and radiologic findings, a presumptive diagnosis of AML and an aneurysm of the right kidney was made.

A right radical nephrectomy using transperitoneal approach was performed with the following findings: adrenal gland with a mass adhered to the upper pole of right kidney and liver (Fig. 2G-I); histopathology examination confirmed the diagnosis of AML. The post-surgical period was uneventful with preserved renal function, but haemoglobin of 7.9 mg/dl was identified, and the patient received a transfusion of a packed red cell with no complications. The patient was discharged five days after surgery with routine immediate post-surgical follow-up.

Discussion.

Angiomyolipoma is the most common mes-

enchymal renal neoplasm, is observed in 0.1 to 0.22% of the general population and is four times more common in female, they are commonly found in the kidney in clinical practice [1].

We consider this report relevant contemplating four elements. First, the clinical presentation is unusual as most of the patient's debut with flank pain and hematuria; in our case, it was associated with diaphoresis, fainting and syncope also the patient referred a non-quantified weight loss with progressive fatigue, which are not typical symptoms of sporadic AML.

Second, although CT is the investigation of choice because of its high sensitivity for detecting fat; usually, most aneurysms are invisible on contrast-enhanced CT, and aneurysm (as described in the present case) is not common [4, 6]. It was possible to observe the intratumoral aneurysm in the 3D- CT reconstruction.

Third, we did not expect an aneurysm because it is associated with tuberous sclerosis and the patient did not show evidence of this diagnosis. The aneurysm found in this patient had 28 mm of a diameter which was above the 95 per-

tile reported by Yamakado et al. [9]. He found that in patients with angiomyolipoma, aneurysms with a mean size of 13.3 mm $\pm$ 6.2 mm belonged to the group of increased risk ruptured aneurysm which contrasts with aneurysms with a mean of 2.4 mm $\pm$ 2.9 mm that corresponded to the unruptured group. Angiomyolipomas $\geq$ 4 cm and aneurysms $\geq$ 5 mm have been used as predictors of rupture; with a sensitivity and specificity of 100% and 38% respectively [9].

Fourth, the two-step treatment, in this case, transarterial embolisation of the tumour and aneurysm followed by nephrectomy has been reported scarcely in the literature [10]. Although some authors like Oesterling et al. recommended that symptomatic tumours larger than 4 cm were treated by selective angiographic embolisation or partial nephrectomy; for this case we perform embolisation of the angiomyolipoma and the aneurysm before radical nephrectomy as a way to reduce tumour size, and decrease the risk of bleed-

ing and rupture [11, 12]. Previous reports have revealed how prophylactic embolisation decrease tumour size; this procedure is recommended in high-risk patients with symptomatic tumours, patients with difficult follow up and in women who are planning future pregnancy [13].

Conclusion.

The evaluation of renal masses using CT, 3D reconstruction might help in the visualisation of intratumoral aneurysms. In the suspicion of an AML, clinicians should discard the presence of tuberous sclerosis. Two measures are essential to remember: tumour size $\geq$ 4 cm and aneurysm size $\geq$ 5 mm, as they are predictors of rupture. When nephrectomy is not performed, transarterial embolisation is followed by serial CT or ultrasound during at least one year.

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