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# Reninoma: Radiological Approach for a Rare Kidney Tumor (Case Report)

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**Abstract** **Case Report**

Reninoma (juxtaglomerular cell tumor) is a rare and benign kidney tumor responsible for renin–mediated hypertension. We report a case of severe hypertension in a female child. Contrast-mediated CT scan especially in the delayed phase was the key of diagnosis. Different elements helped with the diagnostic approach, including clinical and biological data, especially severe hypertension and elevated blood levels of renin and aldosterone. Histological examination after tumor enucleation confirmed the diagnosis.

**Keywords:** Reninoma, blood hypertension, Computed tomography.



## INTRODUCTION

Reninoma is a rare kidney tumor. It is a cause of renin-mediated hypertension described for the first time in 1967 by Robertson. This benign tumor is usually diagnosed in adolescents and young adults with occasional reports in younger children.

It is a tumor secreting renin and responsible for arterial hypertension by secondary hyperaldosteronism.

Diagnostic imaging helps identifying the origin of excessive renin production and contrast CT scan or MRI represent the modalities of choice. The tumor is treated surgically.

In this paper, we present the case of reninoma in an adolescent girl emphasizing clinical presentation, diagnostic evaluation, medical and surgical treatment of this rare tumor.

## OBSERVATION

Female patient aged 13, presented since two months with polyuric polydipsic syndrome, lethargy, anorexia without fever.

Clinically, she had severe hypertension 21/11 cmHg, tachycardia, urine dipstick analysis found proteinuria and hematuria are detected on urine.

Hypertension normalized after bitherapy including conversion enzyme inhibitor and calcium inhibitors.

In the diagnostic workup of hypertension, echocardiography showed features of hypertensive heart disease and progression towards heart failure.

An abdominal Computed tomographic (CT) is performed to exclude adrenal lesions especially pheochromocytoma. CT eliminated adrenal abnormalities and renal arteries stenosis, but revealed a rounded 25 mm diameter subcapsular mass, 5 mm beneath the capsule, under the surface of the anterior midpole of the right kidney, spontaneously hypodense (25 UH), showing contrast enhancement in the delayed phase (75 UH), although it remains hypodense compared to renal parenchyma (Fig 1).

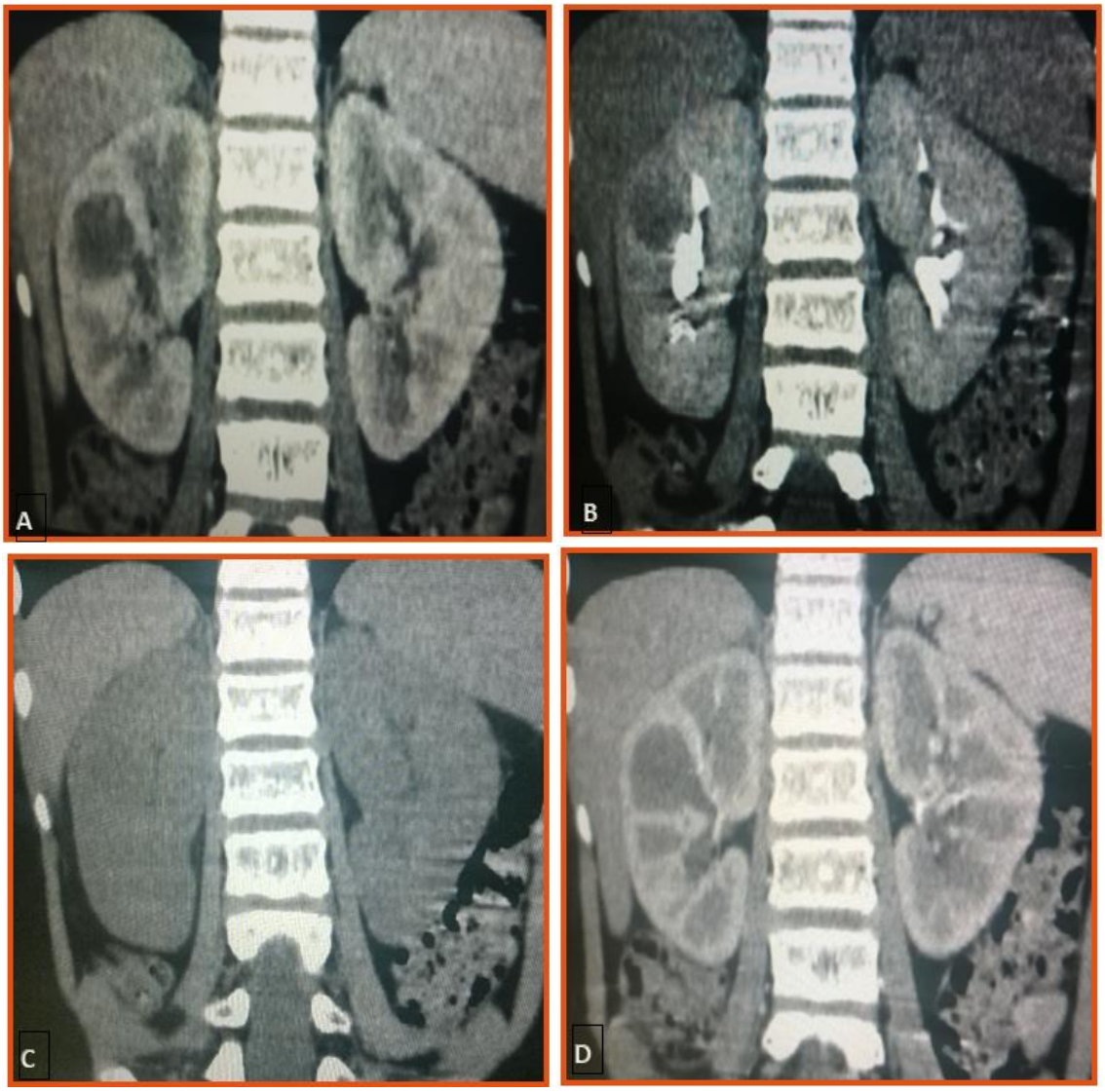


**Fig 1: Abdominal CT scan before (A) and after contrast medium administration at arterial (B), portal (C) and delayed phases (D) Rounded subcapsular mass under the surface of the anterior midpole of the right kidney, corticosinusal, hypodense, showing mild delayed contrast enhancement**

In the ultrasound complement it corresponds to a well delineated, hyperechoic medio-renal mass of the right kidney that demonstrates slight vascularization on Doppler color US (Fig 2).

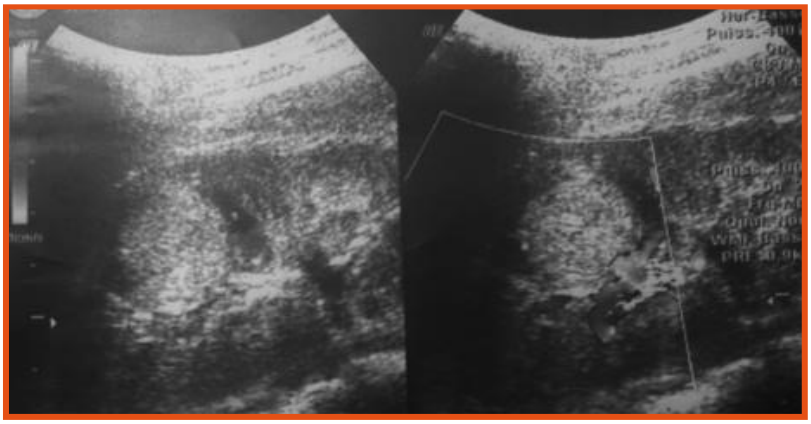

**Fig 2: Longitudinal sonogram of the right kidney: hyperechoic, medio-renal mass showing slight vascularization on doppler color US**

Blood tests showed elevated reninemia (8170mUI/l that represents 240 times the normal) and elevetad aldosteronemia (4045pmol/l that represent 10 times the normal)

All cilnical, biological and radiological data point to a renine producing tumor especially reninoma. The tumor was enucleated and the diagnosis of reninoma was confirmed after pathology examination.

Blood pressure surprisingly normalized after surgery, in the first postoperative day and follow-up was unremarkable.

## DISCUSSION

Reninoma (juxtaglomerular cell tumor) is a benign kidney tumor. It represents a rare cause of renin-mediated hypertension. Since the first report by Robertson *et al*., in 1967 [1], approximately 100 cases of reninoma have been published [2, 3]. These tumors originate from the modified smooth muscle cells of the afferent arteriole of the juxtaglomerular apparatus [2-4].

### Histo Pathology

Histological examination of the excised tumor is essential for confirming the diagnosis of reninoma because other renal tumors can also secrete renin [3]. The correct diagnosis is based on the combination of macroscopic appearance of the tumor, light microscopy of the tumor cells, immune-histochemical staining for various cell markers (including renin), and typical ultrastructural finding of intracellular renin deposits [4].

Reninomas are usually small sub-capsular tumors, well circumscribed, surrounded by a fibrous capsule. Macroscopically, the surface of the tumor is pale or purple with occasional cystic or hemorrhagic areas, with a normal adjacent parenchyma. Microscopically, reninomas comprise closely packed, uniform, round to polyhedral cells with granular, eosinophilic cytoplasm. The nuclei are round to oval with few mitoses. Prominent thick and thin-walled vessels are usually present [5, 6].

The stroma is strewn with mast cells, representing up to 25% of the cells. Mast cells do not exist in the normal juxtaglomerular apparatus. Renin production is demonstrated by in situ hybridization using specific probes labeled by immunofluorescence. In immunohistochemistry, one finds a positivity for antibodies antiactin, antirenin, antiprorenin and anti-CD34. The diagnosis is confirmed by electron microscopy which finds rhomboid-shaped secretion granules [7-9].

### Clinical Presentation

Reninoma is typically diagnosed in adolescents and young adults and the medium of age is 24.4 years, however, occasional cases are reported in younger children [4, 5]. There is a mild female to male predominance with a ratio 2/1.

Patients with reninoma usually present with severe hypertension that is around 198/130 mmhg, revealed by a long history of symptoms such as long-standing history of headaches, malaise, or failure to thrive in younger children; unusually, two cases have been reported without hypertension [6-9]. In fact, the severity of hypertension is not correlated with tumor size [10].

As symptoms are non-specific, there is often a delay in diagnosis of hypertension; however, new imaging techniques have made possible an early diagnosis of the tumor, ranging from 6 years after hypertension detection in the 1991 to 3.5 years recently [6, 12].

We found constantly a hyperreninemia ranging from 3 to 100 times the normal concentration associated to secondary hyperaldosteronemia [13].

### Imaging

Imaging is important for exploring renine mediated hypertension. Ultrasound (US) examination is the first step of the diagnostic workup; it may show an isoechoic, hypo echoic or hyperechoic mass (60% of all cases) although we get some difficulties when the tumor is isoechoic, measuring less than 2 cm [14-16]. Despite these advantages, US is an operator-dependent technology and limited by air-filled bowels and obesity.

The Doppler interrogation of the renal vasculature is indicated to investigate secondary hyperaldosteronemia in the search for renal arteries stenosis.

Computed tomography is considered the mainstay technique in the imaging evaluation of renin-mediated hypertension with 100% sensibility [17]. It shows a small size mass (average 2.5 cm), isodense or hypodense compared to the renal parenchyma; it enhances slightly after contrast administration but remains hypodense to the renal parenchyma.

Sometimes, it is visible only in the delayed phase [18]. In a study, tumor is not detected in a first CT scan performed without delayed phase and diagnosis was established on MRI; the lesion was then viewed after performing an excretory phase on a second CT examination [19].

MRI is indicated when the tumor is clinically suspected but imaging is normal. Per Wang, Rossier and Tanabe, the tumor appears isointense on T1 weighted image compared to normal parenchyma, hypointense on T1 after T1-gadolinium injected sequence with T2

hypointense peripheral halo, although the tumor signal is not reproductive [18, 19].

**Differential Diagnosis**

The first diagnostic challenge is represented by renal tumors secreting renin, especially nephroblastoma, renal cell carcinoma (RCC) and mesoblastic nephroma; the diagnosis is established using clinical, biological and imaging data.

The age range of nephroblastoma is 1 to 5 years, RCC is more frequent in adults but could happen in childhood while mesoblastic nephroma (Bolande tumor) is seen in newborns and infants. All these tumors secret the prorenin which is a renin precursor. Secondary hyperaldosteronemia is caused exclusively by renin secreting tumors which is reninoma.

Imaging characteristics of reninoma are the small size, the homogenous character and hypodense compared to renal parenchyma, unlike other tumors which characterized by bigger size and heterogenous enhancement after contrast administration, especially for nephroblastoma.

**Treatment**

Medical treatment for hypertension is held first then comes surgical procedures.

Surgical resection of reninoma is curative and restores normal blood pressure for the majority of patients. It consists of enucleation with an adequate rim of normal kidney tissue or sometimes by partial nephrectomy. Radiofrequency ablation has been used in two cases as reported in literature [20, 21].

## CONCLUSION

Reninoma is a rare cause of secondary hypertension in young patients. The role of imaging is crucial in investigation and CT scan with delayed post-contrast phase is the modality of choice.

Surgical resection of reninoma is the treatment of choice and leads to normalization of blood pressure. Histopathology confirms diagnosis.

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