

RADIOLOGIC PATHOLOGIC CORRELATION

# Juxtaglomerular Cell Tumor: A Distinct Mesenchymal Tumor of Kidney

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## ABSTRACT

Juxtaglomerular cell tumor (JGCT) is an unusual mesenchymal entity of the kidney. It is a benign renin-secreting tumor causing hypertension and hypokalemia due to secondary hyperaldosteronism. It is curable if it is discovered early and surgically removed, but may cause a fatal outcome usually due to complications of associated hypertension.

**Key words:** Juxtaglomerular cell, kidney, renin

## INTRODUCTION

Juxtaglomerular cell tumor (JGCT), also known as reninoma, is an extremely rare kidney tumor of the juxtaglomerular cells that typically secrete renin. It often causes severe hypertension that is difficult to control in young adults. It was first described, in 1967, by Robertson et al., and was first named, in 1968, by Kihara et al. Since then, approximately 100 case reports have been published. It is generally considered benign, but its malignant potential is uncertain.<sup>[1]</sup>

We report a case of renin secreting tumor diagnosed after nephrectomy through histological and immunohistochemical investigations.

A 35-year-old woman who suffered from hypertension for 3 years that was poorly controlled by medical treatment with angiotensin converting enzyme (ACE) inhibitors was admitted to our institute with a blood pressure of 140/80 mmHg. Laboratory tests showed hypokalemia with a potassium level of 2.63 mmol/l (normal range 3.6-5 mmol/l).

## RADIOLOGICAL FEATURES

Renal ultrasonography showed a round cortical lesion in the right kidney. It was well-circumscribed with heterogenous echogenicity [Figure 1a]. Abdominal computed tomography revealed a low-density tissue lesion indicating a tumor in the right kidney [Figure 1b]. Thus, an open nephrectomy was arranged.

## PATHOLOGICAL FEATURES

In the pathological examination, the surgical specimen showed a yellow tan color and mid-renal lesion (4 cm in diameter). It was well-circumscribed and bound by a thin fibrous capsule.

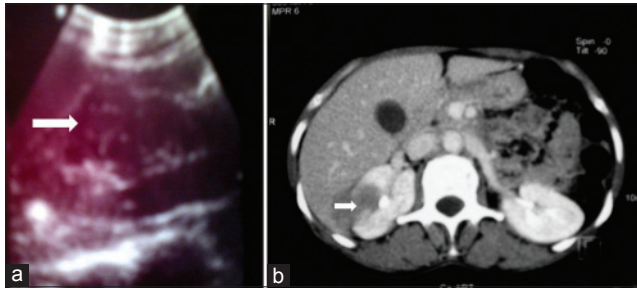
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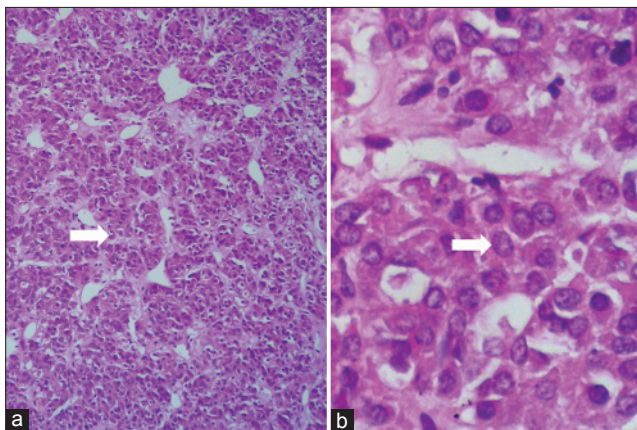
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Microscopically, the tumor consisted of sheets of polygonal and spindle shaped cells with a central regular nucleus. A complex vascular hemangiopericytic pattern and rare tubular elements entrapped were also present [Figure 2].

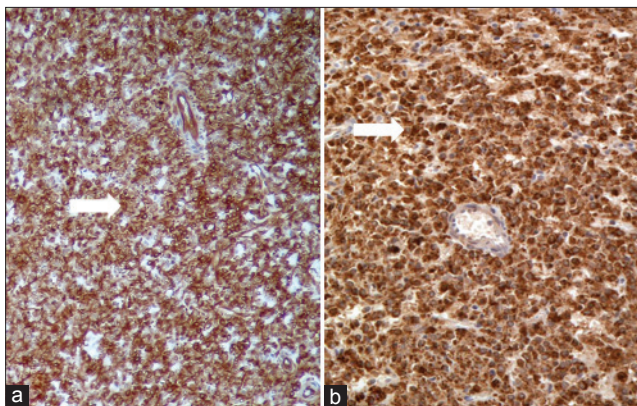
In the immunohistochemical investigation, the tumor cells were found to be immunoreactive for renin, vimentin, and Cluster of Differentiation molecule 34 (CD34) [Figure 3].



**Figure 1:** 35-year-old woman with uncontrolled hypertension diagnosed with juxtaglomerular cell tumor. a) Renal ultrasonography shows a round cortical lesion of the right kidney (arrow). b) Abdominal computed tomography reveals a low-density tissue lesion (arrow) indicating tumor in the right kidney.



**Figure 2:** 35-year-old woman with uncontrolled hypertension diagnosed with juxtaglomerular cell tumor. Hematoxylin and eosin stained tumor tissue - a) at  $\times 10$  magnification shows sheets of polygonal and spindle-shaped cells with a complex vascular hemangiopericytic pattern and b) at  $\times 40$  magnification, shows the tumor cells with eosinophilic cytoplasm and a central regular nucleus.



**Figure 3:** 35-year-old woman with uncontrolled hypertension diagnosed with juxtaglomerular cell tumor. Immunostaining of the tissue demonstrates tumor cells positive for - a) vimentin and b) renin.

These pathological features allowed the diagnosis of JGCT. The postoperative course was uneventful with correction of blood pressure and hypokalemia.

## DISCUSSION

The first example of a JGCT was reported in 1967 by Roberston et al.,<sup>[1]</sup> who described a young male patient with hypertension and a small renal neoplasm. The following year, Kihala et al.,<sup>[1]</sup> reported a similar neoplasm in a young female patient and proposed the term "juxtaglomerular cell tumor".<sup>[2]</sup> Since then, approximately 100 case reports have been published.<sup>[1]</sup> The majority of these patients are women between the ages of 20 and 30 years.<sup>[1,3]</sup>

The diagnosis of JGCT is usually suspected in patients with uncontrolled hypertension, marked hypokalemia, and hyperaldosteronism, although one patient had normal blood pressure.<sup>[2]</sup>

In our case, the symptoms were hypertension with discreet hypokalemia. Tests to determine aldosteronism were not conducted and the final diagnosis was made after surgery based on histological features.

In such a context, radiological studies are often helpful in ruling out other causes of hypertension. Renal arteriography shows the majority of JGCT to be hypervascular and helps to rule out renal artery stenosis.

Computed tomography shows a hypovascular and solid mass in most cases. Renal vein catheterization and selective measurement of renin values may help to detect small tumors.<sup>[2]</sup>

Grossly, JGCT is a solid, well-circumscribed lesion that has a yellow – tan color. The tumor is usually smaller than 3 cm in diameter, but cases ranging from 2 mm to 9 cm have been reported. Histologically, it is made of sheets of polygonal or spindle shaped tumor cells with central round regular nuclei, distinct cell borders, and abundant granular eosinophilic cytoplasm. Typically, tumors have a complex vascular hemangiopericytic pattern.<sup>[3]</sup> Papillary architecture, microcystic pattern, and prominent tubular elements, either neoplastic or entrapped, are also present.<sup>[3,4]</sup> Marked nuclear atypia and mitotic activity are uncommon.<sup>[1]</sup> It is important to distinguish these morphological features from those seen in glomus tumor, hemangiopericytoma, metanephric adenoma, papillary renal cell carcinoma, collecting duct carcinoma, urothelial carcinoma, renal epithelioid angiomyolipoma, and Wilms' tumor.<sup>[1,5]</sup> In these cases, immunohistochemical investigation and ultrastructural studies using electronic microscopy are necessary.

JGCTs are immunoreactive for renin, actin, vimentin, and CD34. Ultrastructural features include abundant rough endoplasmic reticulum, a well-developed Golgi apparatus, and numerous peripherally located sharply angulated rhomboid renin protogranules.<sup>[3]</sup>

Majority of the cases with JGCT are benign, and neither local recurrence nor metastasis has occurred after either radical or partial nephrectomy. However, a few metastatic cases of JGCT have been reported.<sup>[6,7]</sup> The complete tumor resection by radical or partial nephrectomy is the best modality for JGCTs. Antihypertensive agents should be the treatment for hypertension until accurate diagnosis is made. Blood pressure and plasma renin level usually normalize after nephrectomy.<sup>[8]</sup>

## CONCLUSION

In summary, primary JGCT is a distinct mesenchymal tumor entity distinct from other renal tumors. There are only a few genetic studies of JGCT because of the rarity of this disease. It is a benign tumor, but may affect the vital prognosis due to complications of associated hypertension.<sup>[9]</sup>

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