



## Oncology

# A juxtaglomerular cell tumor revealed by a hemorrhagic stroke. A case report

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## ABSTRACT

With about 110 cases reported in literature, juxtaglomerular cell tumors are rare. We report a 25 years old patient who was admitted in neurology for a hemorrhagic stroke secondary to a cerebral aneurysm rupture due to high blood pressure. Etiological investigations showed a solid mass of the left kidney. A radical nephrectomy was realized and pathological examination and immunohistochemical profile concluded to juxtaglomerular cell tumor. The originality of this observation is based on the mode of presentation of a rare renal tumor by a malignant high blood pressure.

## 1. Introduction

Juxtaglomerular cell tumor (JGCT) is an exceptionally rare condition that primarily occurs in young adults. It is an uncommon cause of hypertension, and so far, the medical literature has documented nearly 100 reported cases. The present study discusses a unique and atypical presentation of this tumor.

## 2. Observation

A 25-year-old male patient, with no medical history was admitted for an abrupt onset of left body side weakness (see Fig. 1). That weakness was preceded by acute headaches. Physical examination found left-sided hemiparesis and blood pressure was 220 × 140 mmHg. A brain CT scan showed a right Sylvian subpial hematoma due to a ruptured cerebral aneurysm. A decompressive craniotomy with clamping of the aneurysm has been performed. Unfortunately, neurologic deficiency of the left side of the body persisted. Etiological investigations showed normal kalemia. The measurement of 24 hours of urine normetanephrine and metanephrines was normal. Abdominal CT showed a 65 mm solid mass developed in the lower pole of the left kidney, with a moderate contrast enhancement, distorting the lower calyx. Adrenal glands were normal. MIBG scintigraphy did not localize any abnormal adrenal tissue. A renal cell carcinoma with paraneoplastic hypertension was strongly suspected. A radical nephrectomy has been performed, with no complications during and after the intervention. The tumor was macroscopically circumscribed with a yellowish cut surface and hemorrhagic foci

(Fig. 2). Histological examination shows a tumor proliferation of uniform polygonal cells with eosinophilic cytoplasm and a clear halo around the nucleus. The latter was round and uniform with fine chromatin (Fig. 3). The vasculature was prominent with a haemangiopericytoma pattern. An immunohistochemical study revealed diffuse staining for CD34 (Fig. 3) and focal staining for smooth muscle actin. Tumor cells were negative for desmin, HMB45, PS100, Chromogranin A, and synaptophysin. In front of the clinical context and the morphological and immunohistochemical aspects, the diagnosis of JGCT was made. Twelve months after surgery, blood pressure was normal with no recurrence of the tumor. We confirmed that the hypertension was caused by JGCT.

## 3. Discussion

JGCT, or renin-secreting tumor or reninoma, is a rare benign neoplasm. About 110 cases have been reported in the literature.<sup>1</sup> Clinically, it may present with headaches, nausea, vomiting, impaired vision, and polyuria.<sup>2</sup> These events are related to high blood pressure (HBP) and hypokalemia. Hypertension is inconstant, but it can be malignant. The tumor was revealed by a hemorrhagic stroke in our patient. A similar case was reported in 1974 and was lethal.<sup>2</sup>

Biologically, hypokalemia is suggestive but inconstant. Hormonal investigations show high plasmatic levels of renin and aldosterone. However, a few cases with normal blood tests have been reported.<sup>3</sup> Serum potassium was normal in our patient.

Histologically, the tumor affects the afferent arterioles of the

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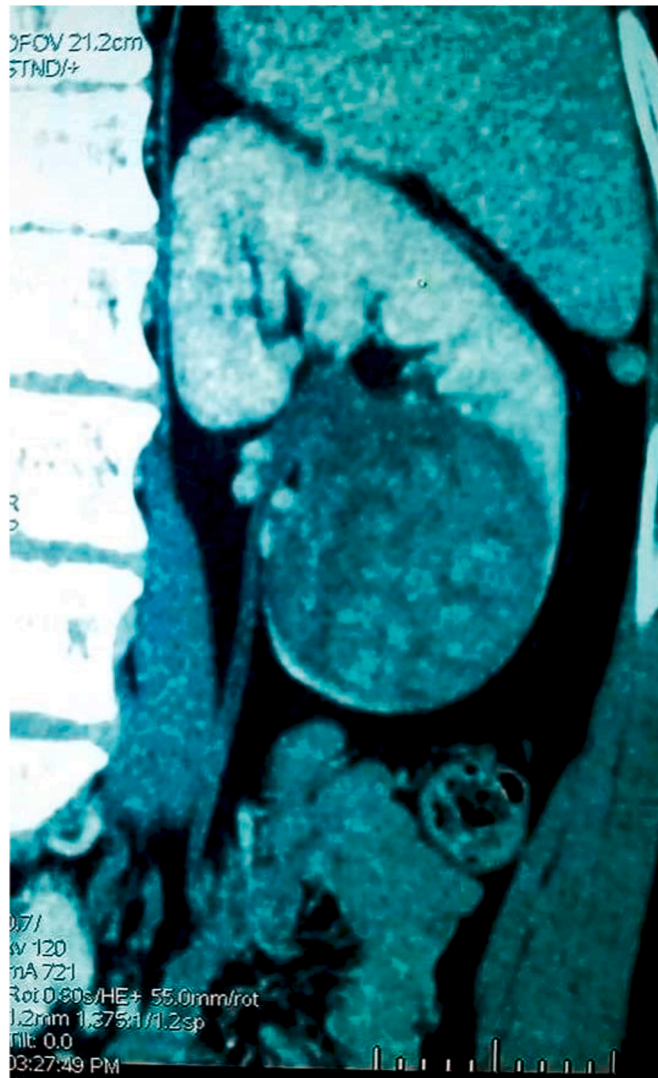


Fig. 1. Coronal section of CT scan showing a left lower pole renal mass, measuring 65 mm.

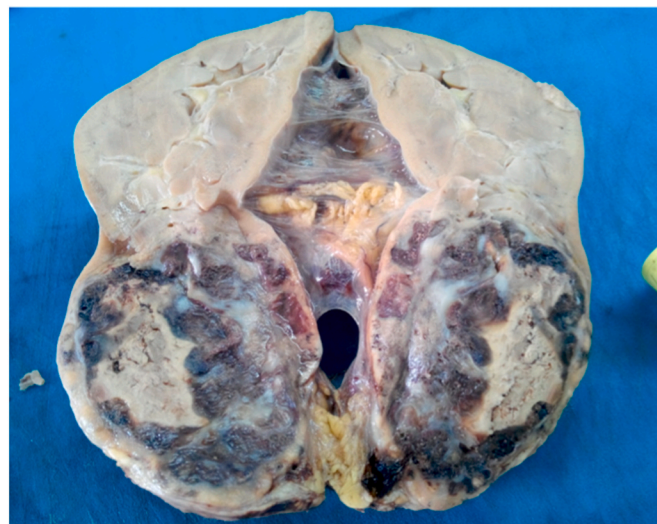
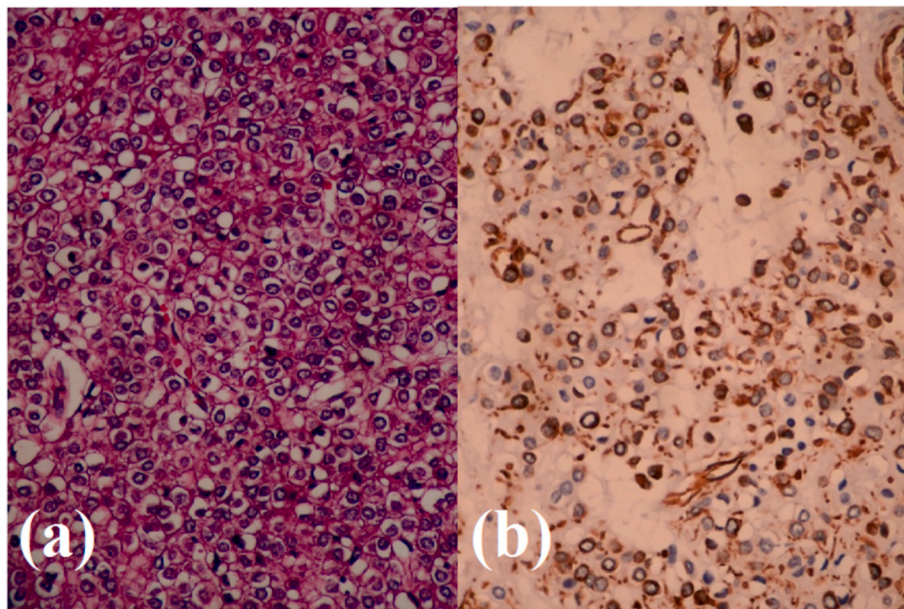


Fig. 2. Macroscopic finding. The tumor is well circumscribed with a yellowish cut surface and hemorrhagic foci.



**Fig. 3.** (a) The tumor consists of uniform polygonal cells with eosinophilic cytoplasm and a clear halo around the nucleus (HE x 200). (b) Intense and diffuse expression of CD34 by tumor cells (CD34 x 200).

glomerulus, but may also be located in the inter-lobular arteries. Cytoplasm is rich in eosinophilic granules. The presence of renin is confirmed after immunohistochemical examination.<sup>4</sup> The histological diagnosis can be difficult when hypertension's clinical context is unknown. The main differential diagnosis is renal cell carcinoma, which can also, in rare cases, be responsible for secondary hypertension and expressed renin. The second major differential diagnosis is solitary fibrous tumor, due to the characteristics of the vascularization and positivity for CD34.

Hypertension usually returns to normal immediately or gradually after tumor removal.

JGCT is considered as a benign tumor, but one case with pulmonary metastasis was reported.<sup>5</sup> Otherwise, the prognosis is especially related to the consequences of HBP, such as in our case. If no complications occurred, HBP is often asymptomatic, so that screening must systematically be done during any medical consultation.

#### 4. Conclusion

Juxtaglomerular cell tumors are rare and benign. However, the consequences of HBP may be serious, especially when it occurs in young

patients. Taking blood pressure during the university medical consultation or pre-hiring consultation could avoid these severe complications.

#### Declaration of competing interest

The authors declare that they have no competing interests.

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