BJMS
www.batnajms.net

Batna Journal of Medical Sciences 2026;13(6):28
https://doi.org/10.48087/BJMS.2026.130628

CASE REPORT

Primary neuroendocrine tumor of the kidney: an exceptional entity illustrated by a case report

Sihem BENSALEM[1], Amina KHODJA[1], Abdelaziz AMMARI[2], Assia BENSALEM[2]

ABSTRACT

Primary renal neuroendocrine tumors (NETs) account for less than 1% of all renal neoplasms and present a significant diagnostic challenge, as their radiological features often mimic those of conventional renal cell carcinoma (RCC). We report the case of a 52-year-old woman who presented with isolated right-sided flank pain. Imaging revealed a 5 cm solid mass in the right kidney, leading to a preoperative diagnosis of RCC and subsequent radical nephrectomy. The definitive diagnosis of a primary renal NET was established based on histopathological examination and a comprehensive immune-histochemical (IHC) panel. The tumor cells demonstrated strong positivity for chromogranin A, synaptophysin, and CD56, while showing no expression of renal epithelial markers such as PAX8. Metastatic workup was negative. Given the unpredictable biological behavior of these tumors, the patient was enrolled in a long-term surveillance program. At three years of follow-up, she remains asymptomatic, with no evidence of disease recurrence. This favorable short- to mid-term outcome is encouraging but highlights the importance of continued close monitoring.

Keywords: Primary renal neuroendocrine tumor, Kidney neoplasm, Immunohistochemistry, Radical nephrectomy, Chromogranin A.

1. Endocrinology-Diabetology and Metabolic diseases, Regional University Military Hospital Commander Abdellali BENBAATOUCHE (HMRUC) Constantine, Faculty of Medicine, University Constantine 3, ALGERIA 2. Medical Oncology Department, Hospital Establishment DIDOUCHE Mourad, Faculty of Medicine, University Constantine 3, ALGERIA

Received: 10 Mar 2026
Accepted: 13 May 2026

Correspondance to: Abdelaziz AMMARI
E-mail: abdelazizammari@yahoo.fr

1. INTRODUCTION

Renal neoplasms are predominantly of epithelial origin, with clear cell renal cell carcinoma (RCC) accounting for over 80% of all malignant kidney tumors. However, the kidney can also give rise to a spectrum of rare neoplasms, including primary neuroendocrine tumors (NETs), which account for less than 1% of all renal tumors [1]. Their pathogenesis remains debated, and their biological behavior is highly unpredictable, ranging from indolent to aggressively metastatic [2]. Clinically and radiologically, they are often indistinguishable from RCC, leading to frequent misdiagnosis [3]. A definitive diagnosis relies exclusively on histopathological and immunohistochemical analysis to confirm neuroendocrine differentiation and exclude a metastasis from a more common primary site. Given their rarity and diagnostic challenges, each documented case provides valuable clinical insight [4].

2. CASE REPORT

This manuscript was prepared in accordance with the CARE (CAse REport) guidelines. As a retrospective analysis, it was exempt from formal ethics committee review, and all patient data were anonymized. A 52-year-old female with no significant past medical history presented to our clinic with a chief complaint of isolated right-sided flank pain. A thorough review of systems was negative for any signs of carcinoid syndrome, such as flushing, diarrhea, or wheezing, and no paraneoplastic syndromes were identified. Her physical examination was notable only for tenderness upon palpation in the right costovertebral angle. While standard laboratory tests, including a complete blood count and comprehensive metabolic panel, were within normal limits, specific neuroendocrine markers

such as serum chromogranin A and urinary 5-hydroxyindolacetic acid (5-HIAA) were not measured pre-operatively due to the low initial suspicion.

An abdominal ultrasound revealed a solid, hypoechoic mass in the upper pole of the right kidney. A subsequent contrast-enhanced computed tomography (CT) scan confirmed a well-circumscribed, 5.0 cm solid renal mass with heterogeneous enhancement and no evidence of regional lymphadenopathy or distant metastases (Figure 1).

Figure 1. Contrast-enhanced computed tomography (CT) scan of the abdomen showing a well-circumscribed, solid, heterogeneously enhancing mass (arrow) measuring 5.0 cm in the upper pole of the right kidney.

Based on these radiological features, the primary pre-operative diagnosis was suspected clear cell RCC. After multidisciplinary discussion, a surgical plan for an open right radical nephrectomy was formulated. Gross examination of the nephrectomy specimen revealed a well-circumscribed, tan-brown mass. Microscopic examination showed a proliferation of neoplastic cells arranged in trabecular and nested patterns, with uniform round nuclei and abundant eosinophilic granular cytoplasm. To determine the tumor's lineage, a comprehensive IHC panel was performed. The neoplastic cells demonstrated strong and diffuse positivity for chromogranin A, synaptophysin, and CD56 (Figure 2).

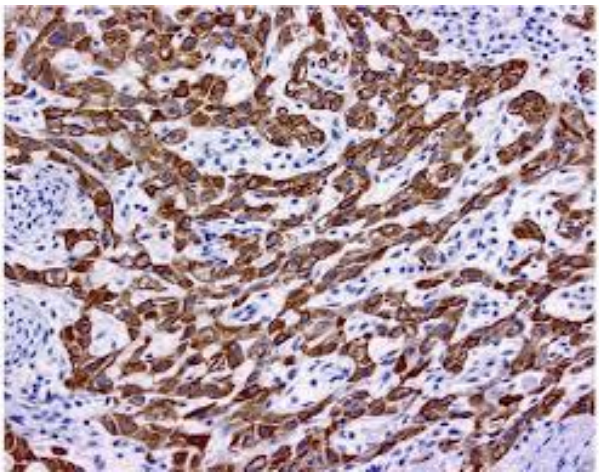

Figure 2. Immunohistochemical staining of the tumor cells. Strong, diffuse cytoplasmic positivity for synaptophysin.

Crucially, the tumor cells were uniformly negative for the renal epithelial markers PAX8 and the RCC marker. This specific immunoprofile established the definitive diagnosis of a primary well-differentiated neuroendocrine tumor of the kidney and effectively excluded a metastatic lesion.

A comprehensive post-operative metastatic workup was negative. The patient was placed on a structured surveillance program, consisting of clinical examinations and contrast-enhanced CT scans of the chest, abdomen, and pelvis every six months for the first two years, and annually thereafter. At the most recent follow-up, three years post-surgery, she remained completely asymptomatic with no evidence of local recurrence or distant metastatic disease.

3. DISCUSSION

The principal finding of this report is the diagnosis and management of a primary renal NET, a rare neoplasm that presents significant challenges. The initial clinical and radiological presentation was non-specific and indistinguishable from conventional RCC, a well-documented diagnostic ambiguity [3]. This highlights that while imaging is essential, it cannot reliably differentiate these entities.

In this context, histopathological and immunohistochemical interrogation becomes indispensable. The definitive diagnosis in our patient was established by a characteristic IHC profile: strong positivity for neuroendocrine markers (chromogranin A, synaptophysin, CD56) coupled with the absence of lineage-specific renal epithelial markers (PAX8, RCC marker). This dual approach is paramount for resolving complex diagnostic dilemmas.

The choice of radical nephrectomy over nephron-sparing surgery (partial nephrectomy) was made pre-operatively based on the radiological appearance and the high suspicion for conventional RCC, where radical surgery is often standard for central tumors. In retrospect, for a 5 cm upper pole tumor, a partial nephrectomy could have been considered to preserve renal function. This case highlights the diagnostic challenge where the pre-operative certainty of RCC guided the surgical extent.

The pathogenesis of primary renal NETs remains debated, and their biological behavior is unpredictable. While our patient's outcome at three years is encouraging, it does not negate the necessity of continued follow-up, as late recurrences have been documented [5]. This inherent uncertainty in prognosis makes long-term surveillance a non-negotiable component of management. The primary limitation is the nature of a single case report, but the accumulation of such cases is essential for building a more complete understanding of this rare entity.

4. CONCLUSION

The management of a primary renal neuroendocrine tumor requires a high index of suspicion and reliance on advanced pathological techniques. A definitive diagnosis is contingent upon a robust immunohistochemical panel. Surgical resection remains the primary therapeutic modality. However, the heterogeneous and unpredictable clinical course of these tumors limits the prognostic value of histology alone. Consequently, a structured, long-term surveillance strategy is essential for the timely detection of recurrence and ensuring optimal long-term patient outcomes. This case reinforces the critical need for a multidisciplinary diagnostic pathway and highlights that ongoing vigilance is non-negotiable in the post-operative care for this rare disease.

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

Funding: This research received no external funding.

REFERENCES

1. Hansel DE, Strosberg JR, Hanks DK. Primary neuroendocrine tumors of the kidney. Semin Diagn Pathol. 2014;31(4):322-327.
2. Moch H, Gasser T, Amin MB, Torhorst J, Sauter G, Mihatsch MJ. Neuroendocrine tumors of the kidney. Virchows Arch. 2003;442(5):449-459.
3. Kim KH, Kim SD, Joo HJ, et al. Primary neuroendocrine tumor of the kidney: radiological and pathological correlation. Korean J Radiol. 2010;11(2):209-214.
4. Gagnier JJ, Kienle G, Altman DG, Moher D, Sox H, Riley D; CARE Group. The CARE guidelines: consensus-based clinical case reporting guideline development. J Med Case Rep. 2013;7:223. doi:10.1186/1752-1947-7-223
5. Partelli S, Berruti A, Rinke A, et al. Long-term outcomes of patients with pancreatic neuroendocrine tumors and liver metastases. Endocr Relat Cancer. 2019;26(6):602-610.