ARCHIVES OF HELLENIC MEDICINE: ISSN 11-05-3992

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Medical Imaging Quiz – Case 84

ARCHIVES OF HELLENIC MEDICINE 2026, 43(3):426–427
ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2026, 43(3):426–427

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A 65-year-old man with a history of hypertension was referred for evaluation of asymptomatic microscopic hematuria detected during a routine examination. Physical assessment was unremarkable, and laboratory tests showed stable renal function. Abdominal magnetic resonance imaging (MRI) revealed a well-circumscribed, hypovascular lesion in the upper pole of the left kidney without lymphadenopathy or metastases. Percutaneous biopsy demonstrated neoplastic cells positive for Vimentin, CD10, P504S, and CK7, and negative for CAIX.

Comments

Papillary renal cell carcinoma (PRCC) represents the second most common histologic subtype of renal cell carcinoma (RCC), accounting for approximately 10–15% of all RCCs. It exhibits distinct morphological, genetic, and clinical characteristics compared to the more prevalent clear cell RCC.

Epidemiology and etiology

PRCC occurs predominantly in males (male-to-female ratio approximately 2:1), with a peak incidence during the fifth to seventh decade of life. It may arise sporadically or as part of hereditary syndromes. The hereditary form, known as hereditary papillary renal carcinoma (HPRC), is associated with activating germline mutations in the MET proto-oncogene located on chromosome 7q31, leading to constitutive activation of the MET tyrosine kinase receptor pathway.

Cytogenetic abnormalities such as trisomy of chromosomes 7 and 17, and occasionally 16 or 20, as well as loss of the Y chromosome in males, are characteristic features. PRCC is also linked to chronic kidney disease and acquired cystic kidney disease, particularly in patients undergoing long-term dialysis.

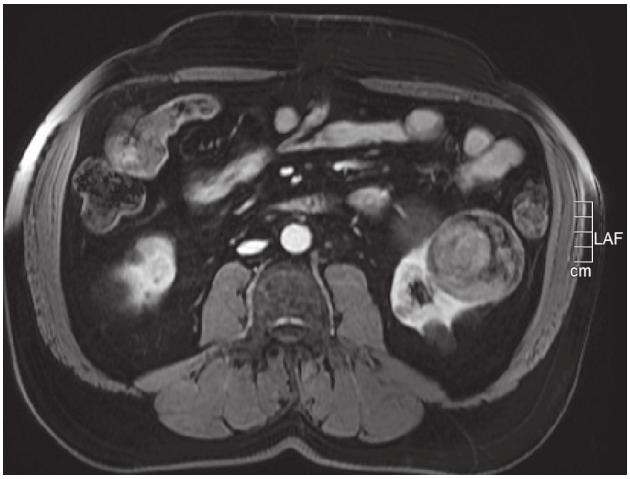

Figure 1. Axial magnetic resonance imaging (MRI) scan of the abdomen demonstrating a mass lesion in the left kidney, consistent with papillary renal cell carcinoma (Fuhrman nuclear grade 1–2), as confirmed by histological examination.

Histopathologic classification

Two main histologic variants of PRCC are recognized: (a) Type 1 PRCC consists of small basophilic cells with scant cytoplasm and low-grade nuclei arranged in papillary or tubular structures. It is frequently associated with MET pathway activation and demonstrates a favorable prognosis. (b) Type 2 PRCC is composed of large eosinophilic cells with pseudostratified, high-grade nuclei and prominent nucleoli. It is genetically more heterogeneous, often harboring CDKN2A silencing, SETD2, or fumarate hydratase (FH) mutations, and is associated with a less favorable clinical outcome.

Morphologic and immunophenotypic features

Macroscopically, PRCC typically presents as a well-circumscribed, solid or cystic renal mass that may be multifocal or bilateral, particularly in hereditary forms. Microscopically, it shows a papillary or tubulopapillary growth pattern with foam cells, psammoma bodies, and frequent intratumoral hemorrhage or necrosis.

Immunohistochemically, PRCC expresses CK7, alpha-methylacyl-CoA racemase (AMACR), CD10, and RCC marker, while Vimentin expression is variable. This immunoprofile assists in differentiating PRCC from other RCC subtypes, such as clear cell or chromophobe carcinoma.

Although there is no absolute rule, in general, papillary renal cancers at diagnosis tend to have a smaller mean diameter and

be at a lower stage. The imaging features of type 1 and type 2 tumors may overlap.

Radiological findings

PRCC generally tends to be hypoechoic on ultrasound.

Whereas according to computed tomography (CT) imaging, smaller lesions tend to be homogeneous in attenuation with larger tumors tending to be heterogeneous. These tumors are characteristically less vascularized than the more common clear cell subtype, showing overall hypoenhancement compared to the adjacent normal renal cortex, particularly seen in the corticomedullary phase. When not enhancing at all, it can be difficult to differentiate from hyper-attenuating cysts 8.

In MRI tumors generally present with a pseudo-capsule. T1: hypointense, T2: hypointense, T1 C+(Gd): Contrast enhancement tends to be less intense than in the more common clear cell RCC subtype, DWI: Restricted diffusion may be useful for differentiating from a hemorrhagic cyst.

Clinical presentation and diagnosis

Most cases are asymptomatic and incidentally detected during imaging for unrelated conditions. When present, symptoms include gross or microscopic hematuria, flank pain, and occasionally a palpable mass. Systemic manifestations such as fever, weight loss, and anemia may occur in advanced disease.

Radiologically, PRCC appears as a hypovascular mass on contrast-enhanced CT or MRI, with less contrast enhancement than clear cell RCC, often displaying central necrosis or hemorrhagic areas.

The definitive diagnosis is established by histopathological evaluation, and staging follows the AJCC TNM classification.

Treatment and prognosis

For localized PRCC, the mainstay of treatment is surgical resection, either by partial nephrectomy (nephron-sparing) or radical nephrectomy, depending on tumor size, location, and renal function.

In metastatic disease, PRCC demonstrates lower sensitivity to conventional vascular endothelial growth factor (VEGF)-targeted therapies (e.g., sunitinib) compared to clear cell RCC. Emerging evidence supports the use of MET inhibitors such as cabozantinib, savolitinib, or crizotinib, and immune checkpoint inhibitors (e.g., nivolumab±cabozantinib). Ongoing clinical trials are investigating combination strategies and molecularly tailored treatments.

The prognosis of PRCC varies by subtype and stage. Type 1 PRCC is generally indolent, with >90% five-year survival in localized disease, whereas type 2 PRCC exhibits more aggressive behavior, with five-year survival rates ranging from 55–70% depending on grade and stage.

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