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The radiologist's role in the management of papillary renal cell carcinoma☆

M.Á. Corral de la Calle[a,*], J. Encinas de la Iglesia[a], M.R. Martín López[b], G.C. Fernández Pérez[c], D.S. Águeda del Bas[a]

[a] *Servicio de Radiodiagnóstico, Complejo Asistencial de Ávila, Ávila, Spain*
[b] *Servicio de Anatomía Patológica, Complejo Asistencial de Ávila, Ávila, Spain*
[c] *Servicio de Radiodiagnóstico, Hospital Universitario del Río Hortega, Valladolid, Spain*

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Abstract Papillary carcinoma is the second most common renal cell carcinoma. It has a better prognosis than the more frequent clear cell carcinoma, although this does not hold true for advanced cases, because no specific treatment exists.

It presents as a circumscribed peripheral tumor (small and homogeneously solid or larger and cystic/hemorrhagic) or as an infiltrating lesion that invades the veins, which has a worse prognosis.

Due to their low vascular density, papillary renal cell carcinomas enhance less than other renal tumors, and this facilitates their characterization. On computed tomography, they might not enhance conclusively, and in these cases they are impossible to distinguish from hyperattenuating cysts. Contrast-enhanced ultrasonography and magnetic resonance imaging are more sensitive for detecting vascularization. Other characteristics include a specific vascular pattern, hypointensity on T2-weighted images, restricted water diffusion, and increased signal intensity in opposed phase images.

We discuss the genetic, histologic, clinical, and radiological aspects of these tumors in which radiologists play a fundamental role in management.

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* Corresponding author.
E-mail addresses: migcorral@gmail.com, migcorral@hotmail.com (M.Á. Corral de la Calle).

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Toma clínica de decisiones

Carcinoma papilar de células renales: el papel del radiólogo en su manejo

Resumen El carcinoma papilar es el segundo renal en frecuencia. Su pronóstico es mejor que el del más frecuente carcinoma de células claras, aunque no en casos avanzados al no existir terapias específicas.

Se presenta como un tumor periférico circunscrito (pequeño y homogéneamente sólido o mayor quístico-hemorrágico) o como una lesión infiltrante e invasora de venas, con peor pronóstico.

Por su baja densidad vascular, el realce es menor que en otras neoplasias renales, lo que facilita su caracterización. En tomografía computarizada puede no realzar de manera concluyente, y entonces es indistinguible de un quiste hiperatenuante. La ecografía con contraste y la resonancia magnética son más sensibles para detectar vascularización. Son además característicos un patrón vascular específico, hipointensidad en T2, restricción de la difusión del agua y aumento de señal en fase opuesta.

Nuestro objetivo es presentar los aspectos genéticos, histológicos, clínicos y radiológicos de estas neoplasias, en cuyo manejo tiene un papel fundamental el radiólogo.

Introduction

Back in 2004, the World Health Organization (WHO) established the actual classification of renal tumors that would modify the former Mainz (about morphology, 1986) and Heidelberg (1997)[1] classifications. The growing protagonism of genetic criteria is a rapid evolution of this classification, with constant introduction of new concepts and entities.

Ninety per cent of all adult malignant renal neoplasms are renal cell carcinomas (RCC) that amount to 2–3 per cent of the total.[2] Its diagnosis grows at an annual rate of 2 per cent[3] as a consequence of the progressive use of diagnostic image modalities since the finding of RCC is usually incidental.

The most common type of RCC is the clear cell renal cell carcinoma (CCRCC) formerly known as hypernephroma due to the similarity of its cells of a clear cytoplasm with adrenal cells. The second most common type of RCC is the papillary renal cell carcinoma (PRCC) that would be consistent to the ''eosinophilic'' subtype in Mainz classification that and amounts to 10–20 per cent of all diagnosed RCCs[1,4–6]–64 per cent of all those that are not CCRCC.[7] The fact that the most common epithelial tumor of all in histological studies–the renal adenoma is considered its precursor and considering the hypovascular character of PRCC–which in turn makes image characterization difficult grows suspicion of misdiagnosis. The chromophobe cell renal carcinoma (CCRC) and other subtypes are more rare.

Age and sex distribution of PRCC is similar to that of CCRCC and affects more commonly middle-aged males and elderly adults. The preference toward males is even more significant in the PRCC with ratios of 2–4:1.[6,8,9] A minority of all PRCCs (around 24 per cent) are asymptomatic.[8]

Our goal is to present the genetic, histologic, clinical and radiologic aspects of these neoplasms. In disease management radiologists play a very important role.

PRCC histology, genetics and subtypes

The ultimate trait of a PRCC is its predominant papillary architecture with a fibrovascular central structure (with machrophages and psammoma bodies in the stroma and a poor but organized vascularization) covered by cells with scarce cytoplasm.[1,4]

In subtype#1, the most common of all, (PRCC1, formerly known as ''basophilic''), these cells are cubic in appearance with small and uniform oval nuclei arranged in a monostratified layer (Fig. 1A). These tumors are usually Fuhrman grade 1–2.[6]

In subtype#2 (PRCC2, formerly known as ''eosinophilic'') that approximately amounts to 37.7 per cent of all PRCCs, these cells have larger spherical nuclei and often a higher nuclear index and atypia and they are arranged in a pseudostratified layer (Fig. 1B). They are usually ranked as nuclear grade 3–4.[5,6]

Around 17 per cent do not fall into either one of these two subtypes and are considered undetermined.[5,6]

Some authors suggest that the papillary adenocarcinoma would be a precursor to the PRCC with a similar evolutionary sequence to the one that leads from colorectal adenoma to carcinoma.[2] The adenoma is a milimetric, solid nodule with tubulopapillary architecture and genetic mechanisms similar to those of PRCC that is usually found in the renal cortex (10–40 per cent of specimens).[1,4]

A small percentage of PRCC can display cells with more abundant clear cytoplasm. Also it is not exceptional that other RCCs - CCRCC included, contain relatively extense foci with papillary architecture that in turn can make histological diagnosis difficult to achieve.[6]

The clear cell papillary renal cell carcinoma is an independent entity from the PRCC and the CCRCC with a different genetic makeup too. It is a well-established tumor