Atlas of Genetics and Cytogenetics
in Oncology and Haematology

OPEN ACCESS JOURNAL AT INIST-CNRS

Solid Tumour Section

Mini Review

Kidney: Papillary renal cell carcinoma

Jérôme Couturier

Department of Pathology, Institut Curie, Paris, France

Published in Atlas Database: May 1998

Online updated version: http://AtlasGeneticsOncology.org/Tumors/kidney5003.html
DOI: 10.4267/2042/37465

Identity

Other names: Chromophilic renal cell carcinoma

Classification

Note: Renal cell carcinomas form a heterogeneous group of tumours which can be divided into several subgroups according to their cytology, architecture, and the part of the nephron from which tumour cells are derived: five basic cell types are recognized: clear-cell, chromophilic-cell (also called papillary renal cell carcinomas (PRCC), herein described), chromophobic-cell, oncocytic-cell, and collector duct cell types.
A certain correlation exists between cell type and architecture of the tumour: the clear-cell type tends to show compact growth, while the chromophilic-cell type is more readily associated with tubulo-papillary architecture.

Clinics and pathology

Etiology

A hereditary form, called Hereditary papillary renal cell carcinoma, of autosomal dominant transmission, has been recently recognized; it predisposes to develop multiple, bilateral renal tumours.

Epidemiology

Papillary renal cell carcinomas (PRCC) represent about 10% of all renal cell tumours; there is a clear excess of male patients (male to female ratio: 5 to 1).

Pathology

Papillary renal tumours are composed of at least 50% of papillary structures, formed by connective tissue stalks covered by small or medium size cuboid cells with eosinophilic or basophilic granular cytoplasm.
Renal cortical adenomas are frequently associated with PRCC in the same kidney, suggesting the possibility of transformation from adenoma to carcinoma.

Cytogenetics

Note: Except a very rare subtype characterized by a translocation t(X;1)(p11;q21) or other abnormalities involving Xp11, PRCC do not show a recurrent structural chromosome rearrangement, but display non random numerical abnormalities; this contrasts with clear-cell renal cell carcinomas which are associated with various deletions of 3p.
PRCC are characterized by loss of Y chromosome in men, trisomy or tetrasomy 7 and 17, trisomies 16, 20, and 12, by order of frequency; these chromosome imbalances have been confirmed by genomic comparative hybridization.
Multifocal PRCC show the same numerical anomalies, with trisomies sometimes in various combinations in tumours within the same kidney or in both kidneys.
The confrontation of karyotypic abnormalities with histopathological data suggest that specific chromosome imbalances may make it possible to distinguish benign papillary adenoma from malignant PRCC; the small papillary tumours without any morphological or clinical sign of malignancy are characterized by a combination of loss of Y, and excess of chromosomes 7 and 17, only, whereas tumours with aggressive growth and metastatic dissemination show more complex karyotypic changes, in addition to polysomy 7 and 17; it is suggested that tumours with tri- or tetrasomy 7 and 17 correspond to papillary adenomas, and that tumours with more complex karyotypic changes are papillary carcinomas, irrespective of their size.

Genes involved and Proteins

Note: In hereditary PRCC, linkages studies have shown that the gene of the disease could be located at 7q31.1-34, between markers D7S496 and D7S1837, an interval containing the MET proto-oncogene; missense mutations in the tyrosine kinase domain of the MET

gene have been identified in the germline of affected members of PRCC families, and in a subset of sporadic PRCC.

References

Thoenes W, Störkel S, Rumpelt HJ. Histopathology and classification of renal cell tumors (adenomas, oncocytomas and carcinomas). The basic cytological and histopathological elements, and their use for diagnosis. Pathol Res Pract. 1986 May;181(2):125-43.

Kovacs G. Papillary renal cell carcinoma. A morphologic and cytogenetic study of 11 cases. Am J Pathol. 1989 Jan;134(1):27-34.

Kovacs G, Fuzesi L, Emanuel A, Kung HF. Cytogenetics of papillary renal cell tumors. Genes Chromosomes Cancer. 1991 Jul;3(4):249-55.

Hughson MD, Johnson LD, Silva FG, Kovacs G. Nonpapillary and papillary renal cell carcinoma: a cytogenetic and phenotypic study. Mod Pathol. 1993 Jul;6(4):449-56.

Kovacz G. Molecular cytogenetics of renal cell tumors. Adv Cancer Res. 1993;62:89-124.

Bentz M, Bergerheim US, Li C, Joos S, Werner CA, Baudis M, Gnarra J, Merino MJ, Zbar B, Linehan WM, Lichter P. Chromosome imbalances in papillary renal cell carcinoma and first cytogenetical data of familial cases analysed by comparative genomic hybridization. Cytogenet Cell Genet. 1996;75(1):17-21.

Dal cin P, van Poppel H, van Damme B, Baert L, Van den Berghe H. Cytogenetic investigation of synchronous bilateral renal tumors. Cancer Genet Cytogenet. 1996 Jul 1;89(1):57-60.

Schmidt L, Duh FM, Chen F, et al. Germline and somatic mutations in the tyrosine kinase domain of the MET proto-oncogene in papillary renal carcinomas. Nat Genet. 1997 May;16(1):68-73.

Dal Cin P, Stas M, Sciot R, De Wever I, Van Damme B, Van den Berghe H. Translocation (X;1) reveals metastasis 31 years after renal cell carcinoma. Cancer Genet Cytogenet. 1998 Feb;101(1):58-61.

This article should be referenced as such:

Couturier J. Kidney: Papillary renal cell carcinoma. Atlas Genet Cytogenet Oncol Haematol.1998;2(4):146-147.