Tumors of the kidney



Tumors of the kidney

Types

Benign

Malignant

Childhood tumors

WHO classification of tumours of the kidney

Renal cell turnours		Mesenchymal tumours occurring mainly in ad	ults
Clear cell renal cell carcinoma	8310/3	Leiomyosarcoma	8
Multilocular cystic renal neoplasm of low		Angiosarcoma	9
malignant potential	8316/1*	Rhabdomyosarcoma	8
Papillary renal cell carcinoma	8260/3	Osteosarcoma	Ş
Hereditary leiomyomatosis and renal cell		Synovial sarcoma	ę
carcinoma-associated renal cell carcinoma	8311/3*	Ewing sarcoma	ç
Chromophobe renal cell carcinoma	8317/3	Angiomyolipoma	8
Collecting duct carcinoma	8319/3	Epithelioid angiomyolipoma	8
Renal medullary carcinoma	8510/3*	Leiomyoma	ξ
MiT family translocation renal cell carcinomas	8311/3*	Haemangioma	9
Succinate dehydrogenase-deficient		Lymphangioma	9
renal carcinoma	8311/3	Haemangioblastoma	S
Mucinous tubular and spindle cell carcinoma	8480/3*	Juxtaglomerular cell tumour	8
Tubulocystic renal cell carcinoma	8316/3*	Renomedullary interstitial cell tumour	8
Acquired cystic disease-associated renal		Schwannoma	ç
cell carcinoma	8316/3	Solitary fibrous tumour	8
Clear cell papillary renal cell carcinoma	8323/1		
Renal cell carcinoma, unclassified	8312/3	Mixed epithelial and stromal turnour family	
Papillary adenoma	8260/0	Cystic nephroma	8
Oncocytoma	8290/0	Mixed epithelial and stromal tumour	8
Metanephric tumours		Neuroendocrine tumours	
Metanephric adenoma	8325/0	Well-differentiated neuroendocrine tumour	8
Metanephric adenofibroma	9013/0	Large cell neuroendocrine carcinoma	8
Metanephric stromal tumour	8935/1	Small cell neuroendocrine carcinoma	8
		Phaeochromocytoma	8
Nephroblastic and cystic tumours occurring			
mainly in children		Miscellaneous tumours	
Nephrogenic rests		Renal haematopoietic neoplasms	
Nephroblastoma	8960/3	Germ cell tumours	
Cystic partially differentiated nephroblastoma	8959/1		
Paediatric cystic nephroma	8959/0	Metastatic tumours	
Mesenchymal tumours			
		The morphology codes are from the International Classificatio	n of l
Mesenchymal tumours occurring mainly in children		for Oncology (ICD-O) {917A}. Behaviour is coded /0 for benig	n tur
Clear cell sarcoma	8964/3	/1 for unspecified, borderline, or uncertain behaviour; /2 for ca	arcin
Rhabdoid tumour	8963/3	situ and grade III intraepithelial neoplasia; and /3 for malignar	it tun
Congenital mesoblastic nephroma	8960/1	The classification is modified from the previous WHO classific	ation
Ossifying renal tumour of infancy	8967/0	taking into account changes in our understanding of these les	ions

myosarcoma 8890/3 iosarcoma 9120/3 bdomyosarcoma 8900/3 eosarcoma 9180/3 ovial sarcoma 9040/3 ng sarcoma 9364/3 iomyolipoma 8860/0 helioid angiomyolipoma 8860/1* 8890/0 myoma 9120/0 mangioma phangioma 9170/0 mangioblastoma 9161/1 aglomerular cell tumour 8361/0 omedullary interstitial cell tumour 8966/0 wannoma 9560/0 ary fibrous tumour 8815/1 ed epithelial and stromal tumour family tic nephroma 8959/0 ed epithelial and stromal tumour 8959/0 roendocrine tumours -differentiated neuroendocrine tumour 8240/3 e cell neuroendocrine carcinoma 8013/3 8041/3 all cell neuroendocrine carcinoma eochromocytoma 8700/0 cellaneous tumours al haematopoietic neoplasms m cell tumours astatic tumours horphology codes are from the International Classification of Diseases ncology (ICD-O) [917A]. Behaviour is coded /0 for benign turnours;

unspecified, borderline, or uncertain behaviour; /2 for carcinoma in nd grade III intraepithelial neoplasia; and /3 for malignant tumours. classification is modified from the previous WHO classification {756A}, g into account changes in our understanding of these lesions. *New code approved by the IARC/WHO Committee for ICD-O.

Clear cell renal carcinoma in the focus







(Renal cell carcinoma- RCC, ((Hypernephroma))



RCC - "Classic" symptoms

Haematuria

Hypertension

Costovertebral pain (flank) pain

No symptoms

Most cases are incidental

Diagnostics

US

CT

MR

Preoperative : FNAB

Core-biopsy

Male, 44 y/o A Incidentally found renal tumor

1

MF 1.90

R

TP 262.5 OPTIRAY (kesoi) SPI 5 DR.KOPCSANYI/GJ















Core needle bx Kidney



Characteristics of renal cell cancer

- Clear "plant-like" cells
- Widespread paraneoplasticus signs
- Blood vessel invasion
- Widespread metastases,

anytime,

anywhere (reg. Ln-s, adrenals,lung, brain, bones, thyroid, skin, testis...)



Paraneoplastic signs:

Polycythemia Hypercalcaemia Hypertension Hepatic dysfunction Feminisation/Masculinisation Cushing syndrome Eosinophily Leukemoid reaction Amyloidosis











512*512 MAG*1.9 SH*4

₩180 -65

58

Female, 67 y/o

Easily bleeding polypoid mass in the nasal cavity

Kl.:Haemangioma?









Male, 66 y/o pathologic fracture of the femur, 11 years past nephrectoy for RCC







Male, 64 y/o Left sided stiffness on the neck



DR 60

MI:1.2

2DG 99

ξ_-(

TOSHIBA

Dg.: Most probably RCC







Male, 65 y/o. Pathologic fracture of the humerus. 3 weeks after polster....





Male, 72 y/o. 4.5 cm-sized cold nodule in the thyroid. A.: neg.








Risk factors

Enviromental effects smoking petrol painting industry heavy metals Laboratory workers asbestosfiremen

obesity hypertension oestrogen therapy Von Hippel Lindau



Most frequent chromosomal abnormalities

Chr. Abn.: 3-3p14/3p.26 trans., del, VHL gene(3p25.3) 7,16,17 trisomy, Yloss, X;1 transl (papill. Cc.) 96% sporadic 4% familiary VHL Hereditary familiar RCC - VHL gene, without any other manifestation of VHL Hereditary papillary cancer



Tumors of the kidney

Histogenesis of renal tumors

Tubular epithelium Mesenchyme Metanephrogenic elements Papillary adenoma Metanephric adenoma Angiomyolipoma Nephroblastoma (Wilms tumor) Oncocytoma Clear cell renal cell carcinoma (renal cell carcinoma: RCC) Multilocular cystic neoplasm Papillary RCC Chromophobe RCC Clear cell papillary RCC

Tumors of the kidney

Benign

Nephrogenic adenoma Oncocytoma Angiomyolipoma Malignanat RCC conventional cystic papillary chromophob cell sarcomatoid UCC Wilms Malignant, non RCC Mesenchymal Lymphoma, leukaemia Mal. angiomyolipoma Metastatic

VI. Tumors of the kidney

Malignant tumors

1. Clear cell RCC

Most common type (approx. 80%)

Characteristic golden yellow color



- Variable architecture: solid, tubular, papillary, microcystic, cystic
- Haemorrhages and necroses are usual
- Genetic/epigenetic alteration: 3p deletion, VHL mutation, VHL hypermetilation









Malignant tumors

Multilocular cystic renal neoplasm of low malignant potential 2-3% of RCCs

Middle-aged patients, usually incidentally detected

Complex cystic lesion by radiological examination

Composed exclusively of thin-walled cysts

Clear cyst content

Low-grade tumor cells: internal surface of cysts, small groups in the septums

Excellent prognosis

Papillary RCC

Second most common (approx. 12-15%)

More commonly multifocal

Type 1

Grey-white, well-circumscribed, encapsulated, haemorrhages Papillary, tubular, or solid architecture

Cuboidal cells, foamy macrophages, psammoma bodies





Papillary RCC

Second most common (approx. 12-15%)

More commonly multifocal

Type 2

Variable appearance, necroses, haemorrhages Larger cells, higher grade, worse prognosis Characteristic genetic alteration: trisomies (**7**, **17**, 12, 16 20), loss of Y, c-Met mutation









Chromophobe RCC Approx. 4-5% Well-circumscribed, grey-white Clear or eosinophilic cytoplasm, distinct cell borders, binucleated figures, rasinoid nuclei, perinuclear halo Widespread chromosomal losses Better prognosis DD: oncocytoma



Benign tumors

Benign tumors

1. Papillary adenoma

Papillary tumor with low-grade tumors cells, ≤ 15 mm

Grey-white, round nodule, can be multifocal

Commonly incidental finding

Cuboidal, monomorphic tumor cells, papillary architecture, psammoma bodies

Oncocytoma 5% of renal tumors Machagony brown, characteristic central scar Nested, trabecular architecture, degenerative signs Oncocytic, bland looking cells (large, granular eosinophilic cytoplasm), however, bizarre nuclei, extracapsular infiltration, vascular invasion might be encountered



1367/04 I/1

Oncocytoma

5% of renal tumors

Machagony brown, characteristic central scar Nested, trabecular architecture, degenerative signs





Oncocytic, bland looking cells (large, granular eosinophilic cytoplasm), however, bizarre nuclei, extracapsular infiltration, vascular invasion might be encountered



Angiomyolipoma

PEComa: <u>perivascular e</u>pithelioid <u>c</u>ell tumor

1% of the resected renal tumors, but it is more common (by US detected tumors that are not operated) Well-circumscribed, usually fatty appearance Adipose tissue, thick-walled vessels, smooth muscle HMB45+, Melan A+ (melanocytic markers!!)



Dg.:Angiomyolipoma



Metanephric adenoma

More common in females

Average size is 5 cm

Grey-brown, solid, cystic degeneration may be present

Well-circumscribed

Tubulary, solid, or papillary architecture

No mitoses



Renal pole re: Hist.?



Dg.: Nephrogén adenoma

Prognostic facts

Tumor type



Kuthi L et al. Pathol Oncol Res. 2017 Jul;23(3):689-698

Prognostic factors

Stage

pT:

pT1 – tumor \leq 7 cm in greatest dimension, limited to the kidney (pT1a \leq 4 cm, pT1b >4 cm)

pT2 - tumor >7 cm in greatest dimension, limited to the kidney (pT2a \leq 10 cm, pT2b >10 cm)

pT3 - tumor extends into major veins (renal vein, VCI) or perinephric tissues (ERE)

(pT3a renal vein invasion and/or ERE, pT3b VCI invasion below

diaphragm, pT3c VCI invasion above diaphragm)

pT4 - tumor invades beyond the Gerota fascia (including contiguous extension into the ipsilateral adrenal gland)

Prognostic facts

Table 2Cox regression analysis for cancer-specific survival rates innon-metastatic clear cell RCC

Characteristic	Hazard ratio	CI 95%	p value
Univariate			
ISUP grade	7.50	5.01-11.21	< 0.001
TNM stage	2.54	2.04-3.15	< 0.001
Surgical margin status	2.95	1.57-5.53	< 0.001
Microscopic tumor necrosis	6.74	4.53-10.07	< 0.001
Rhabdoid/sarcomatoid change	5.14	3.39-7.78	< 0.001
Giant tumor cells	3.93	2.51-6.15	< 0.001
Multivariate			
ISUP grade	4.33	2.36-7.95	< 0.001
TNM stage	1.86	1.49-2.33	< 0.001
Surgical margin status	2.61	1.39–5.2	0.003
Microscopic tumor necrosis	1.69	0.93-3.05	0.081
Rhabdoid/sarcomatoid change	0.96	0.57-1.61	0.896
Giant tumor cells	0.67	0.4–1.13	0.139

Kuthi L et al. Pathol Oncol Res

Prognostic facts

Grade:

Fuhrman grade/ISUP grade

Grade 1: Inconspicuous nucleoli at ×400 magnification and basophilic. **Grade** 2: Clearly visible nucleoli at ×400 magnification and eosinophilic. **Grade** 3: Clearly visible nucleoli at ×100 magnification.



Treatment

Partial (pT1 tumors) or radical nephrectomy (open or laparoscopic surgery)

Targeted therapy (metastatic cases): sunitinib,

Diagnostic difficulties

Challenge of the radiologist and pathologist


47 y/o woman

Nephrectomy for RCC 3 years earlier

Multiple hypoechoic lesions in the retroperitoneal region and omentum

Met.?







CD68

Dg.: Adiponecrosis



Female, 65 y/o Resection of the upper pole of the kidney, 20 y-s earlier

CT control: protruding hypoechoic tumor of 3 cm in diam.











63 y/o female

Smoker

Flank pain, hematuria

Ureteroscope: papillary tumor





Dg.: Tu.,High grade UCC Urothelial (Transitional cell) carcinoma- (UCC) -symptoms

Hematuria

Hydronephrosis

Pain

(Chr. aberrations: 9 monosomy, 9p,9q, 17p, 13q, 11p, 14q del.) Male, 60 y/o Anaemia Haematuria









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