Supplementary Table 1

Criteria for histopathological diagnoses of kidney cysts, malignant and benign renal neoplasms

|  |  |
| --- | --- |
| Diagnosis | Criteria for histopathology |
| ADPKD | * Cysts in renal medulla and cortex, lined with a single layer of flattened to cuboidal epithelium
* Parenchyma in between the cysts with varying degree of chronic inflammation, interstitial fibrosis, glomerular sclerosis and tubular atrophy
 |
| ADPKD with hyperplastic foci  | Scattered foci of hyperplastic, sometimes polypoid or even papillary projections into the cyst lumen |
| Clear cell renal cell carcinoma | * Tumor cells with distinct cell membrane and clear cytoplasm due to dissolved lipids and glycogen during histologic processing
* Possibly minor cell populations with eosinophilic cytoplasm
* Diverse architecture with solid sheets or nests, acinar and alveolar patterns, cystic structures
* Network of thin-walled, small blood vessels
* Areas of calcification, metaplastic ossification or fibromyxoid change
 |
| Papillary renal cell carcinoma | * Papillary and tubular structures, rarely solid
* Stalks contain fibrovascular cores commonly infiltrated by variable number of foamy macrophages
 | Type 1: * A single or sometimes double layer of small cells with pale or clear cytoplasm
* Low nuclear grade with round to oval nuclei, inconspicuous nucleoli
 |
| Type 2:* Usually a single layer of typically eosinophilic cells
* Often higher nuclear grade with pseudostratified nuclei
* Macrophages likely to be found close to necrosis
 |
| Papillary adenoma of the kidney | * Tubular, papillary or tubulopapillary architecture
* Low nuclear grade
* Maximum 5 mm in diameter
* Well circumscribed, sometimes inconspicuous fibrous pseudocapsule
* Cuboidal cells with pale to eosinophilic to basophilic cytoplasm
* (type 1)
* Cells with voluminous, eosinophilic cytoplasm (type 2)
* Possibly psammoma bodies and foamy macrophages
 |
| Multilocular cystic renal cell carcinoma | * Fibrous pseudocapsule
* No expansile tumor nodules
* Whole tumor composed of cysts and fibrous septa with aggregates of epithelial cells with clear cytoplasm indistinguishable from grade 1 clear cell carcinoma
* Cyst lining cells are also clear or lightly eosinophilic and build usually a monolayer; multilayered and minute papillary structures occasionally exist
* Sometimes focally calcifications or metaplastic ossification
 |

References

Eble JN, Sauter G, Epstein I, Sesterhenn I: World Health Organisation Classification of Tumours; Pathology & Genetics; Tumours of the Urinary System and Male Genital Organs.

Bostwick G, Cheng L: Urologic Surgical Pathology; Expert Consult - Online and Print, second edition. *Mosby Elsevier* 2008.

Murphy, Grignon, Perlman: Tumors of the Kidney, Bladder and Related Urinary Structures; AFIP Atlas of Tumor Pathology Vol. 1. 2004.