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.