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Clear Cell Papillary Renal Cell Carcinoma

Anna Caliò[1], Diego Segala[2] and Guido Martignoni[1,2]
[1]Department of Diagnostic and Public Health, Section of Pathology, University of Verona, Verona, Italy
[2]Department of Pathology, Pederzoli Hospital, Peschiera del Garda, Italy

Synonyms

Clear cell tubulopapillary renal cell carcinoma

Definition

Indolent renal cell tumor made up of bland clear cells arranged in tubules and papillae with often linear nuclear alignment away from the basement membrane (Massari et al. 2018).

Clinical Features

- **Incidence**
  This neoplasm accounts for 1–4% of renal tumors. Most tumors are incidentally detected as sporadic or arising in end-stage renal disease and von Hippel-Lindau syndrome (Tickoo et al. 2006; Gobbo et al. 2008).
- **Age**
  There is no age predilection.
- **Sex**
  There is no gender predilection.
- **Site**
  There is no site predilection.
- **Treatment**
  Partial nephrectomy and less frequently, radical nephrectomy is the standard treatment.
- **Outcome**
  To date, neither recurrence nor metastasis has been reported (Massari et al. 2018).

Macroscopy

Tumors are often small (usually pT1a), grayish, well-circumscribed and encapsulated with common cystic change (Fig. 1a).

Microscopy

Tubular, papillary, branched glandular, acinar, and cystic patterns are seen in varying proportions. Neoplastic clear cells are cuboidal with round nuclei and inconspicuous nucleoli (ISUP/WHO grade 1 and 2) arranged in a linear fashion away from the basement membrane. Fibrous and/or smooth muscle stroma in varying amounts may be observed (Fig. 1b).

H. V. Krieken (ed.), *Encyclopedia of Pathology*,
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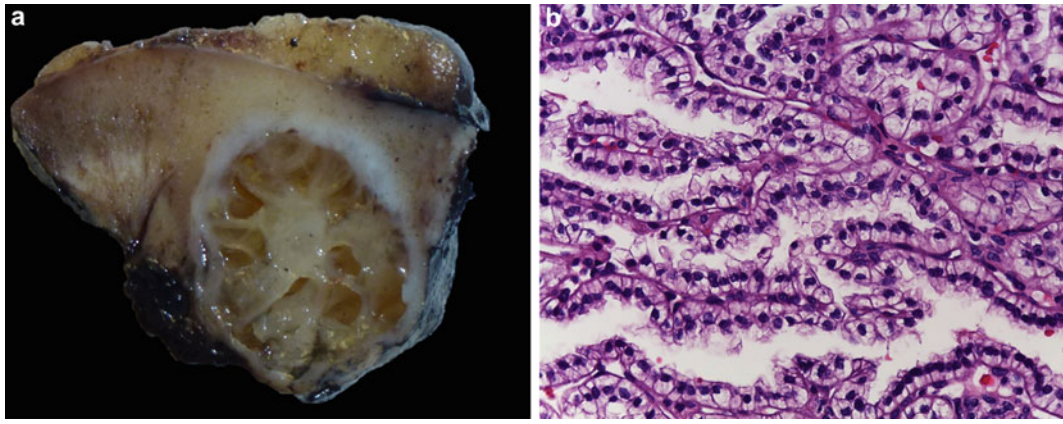

Clear Cell Papillary Renal Cell Carcinoma, Fig. 1 A small, well-circumscribed, and encapsulated with cystic changes mass without the yellowish features typically seen in clear cell renal cell carcinoma (**a**). Cuboidal clear cells with inconspicuous nucleoli in a linear fashion away from the basement membrane arranged in a tubular pattern (**b**)

Immunophenotype

Tumor cells diffusely express cytokeratin7, PAX8, cytokeratin 34βE12, and CAIX (often in cup-like distribution), frequently stain for GATA3; whereas racemase and CD10 are usually negative (Rohan et al. 2011; Martignoni et al. 2017).

Molecular Features

These tumors lack the classic genetic events of clear cell renal cell carcinoma (3p/VHL alteration) and papillary renal cell carcinoma (7 and 17 trisomies) (Rohan et al. 2011).

Differential Diagnosis

The differential diagnosis with clear cell renal cell carcinoma with low nucleolar grade may be extremely difficult. Despite immunophenotype being usually diagnostic, in selected cases, molecular analysis could be necessary to classify the neoplasm. Less frequently, papillary renal cell carcinoma with extensively clear cell changes should be ruled out.

References

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