CASE REPORT Open Access

# Low-grade oncocytic tumour of kidney (CD117-negative, cytokeratin 7-positive)



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## **Abstract**

**Background:** The term low-grade oncocytic tumour of kidney is an emerging entity describing CD117-negative and cytokeratin 7-positive indolent tumors with overlapping morphological features between oncocytic tumors.

**Case presentation:** We present herein the case of a 77-year-old female with a 3.2-cm nodule in mid pole of the left kidney. The tumor was uniformly oncocytic with solid, compact nested and trabecular growth patterns. There was common areas of transition to central zones of stromal edema with marked tumor hypocellularity and growth in cords. Some of these areas had adjacent fresh hemorrhage. Immunohistochemistry showed strong and diffuse expression of cytokeratin 7 and negativity for cKIT/CD117.

**Conclusion:** The proper use of this new diagnostic category avoids labeling such tumors as unclassified renal cell carcinoma – a broad category with different morphologic features and heterogenous prognosis.

Keywords: Carcinoma, Renal cell, Kidney neoplasms, Kidney

## **Background**

In 2019, Trpkov and colleagues described an emerging entity among unclassified renal cell neoplasms. They distinguished a new category of oncocytic tumors that do not fit in existing diagnostic entities (Trpkov et al. 2019; Trpkov and Hes 2019). The term low-grade oncocytic tumour of kidney was suggested to describe CD117-negative (in contrast to expected expression in oncocytomas and chromophobe carcinomas) and cytokeratin 7-positive tumors with

overlapping morphological features between oncocytic tumors (Trpkov et al. 2019; Trpkov and Hes 2019). In the original series of 28 patients, all remained alive with no evidence of progression (median 21 months of follow up) after surgery (Trpkov et al. 2019).

### **Case presentation**

We present herein the case of a 77-year-old female who sought medical assistance due back pain for about 12 months. Abdominal magnetic resonance imaging showed a circumscribed solid tumour in the mid pole of the left kidney (Fig. 1a-b). It was a partially exophytic mass with central area suggestive of necrosis / hypovascularity. A radical nephrectomy was performed.

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**Fig. 1** Low-grade oncocytic tumour of kidney. Abdominal magnetic resonance imaging - coronal plane (**a**) and axial (**b**) shows a well circumscribed tumor in the left kidney (T1-weighted scans)

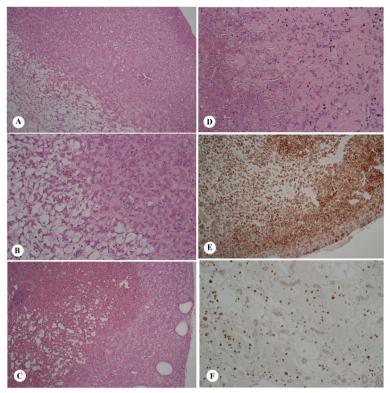
At gross examination, a 3.2-cm brown nodule was observed in mid pole. Cut surface showed central fresh hemorrhage. Tumor was grossly and microscopically confined to the kidney. The tumor cells were uniformly oncocytic with solid, compact nested and trabecular growth patterns. Oncocytic cells shared abundant eosinophilic cytoplasm and nuclei with occasional nucleoli (ISUP grades 1–2) without irregular contours such as "raisinoid"-shaped nuclei of chromophobe carcinomas. These solid areas showed common areas of transition to central zones of stromal edema with marked tumor hypocellularity and growth in

cords (Fig. 2a-b). Some of these areas had adjacent fresh hemorrhage (Fig. 2c-d). Necrosis was not observed. Immunohistochemistry showed strong and diffuse expression of cytokeratin 7 (Fig. 2e) and negativity for cKIT/CD117 (Fig. 2f) in different blocks. Additional reactions showed positivity for PAX8 and absent expression of alpha-methylacyl-CoA racemase.

#### Discussion

The case presented herein fits in the description of this new entity. In the original series of 28 patients by Trpkov and colleagues, median age at diagnosis was 66 years and there was a slight female predominance 1.8: 1. Median tumor size was 3.0 cm and 68% of the tumors measured < 4.0 cm and were staged as pT1a (Trpkov et al. 2019). In addition to the proper immunoprofile, the case presented herein showed the described typical findings of coexistent fresh hemorrhage with transition to edematous stromal and growth pattern in cords.

The main differential diagnoses in this context are other oncocytic tumors of the kidney. Oncocytoma may have a broad spectrum of morphologic features (Trpkov et al. 2010). Oncocytomas are, however, typically CD117-positive and show a particular pattern of cytokeratin 7 expression - either entirely negative or predominantly negative with strong staining of isolated tumor cells or small tumor cell clusters. The feature of fresh hemorrhage and growth in cords intermixed in a edematous stroma (characteristic of low grade oncocytic tumour of kidney) is different from the insular growth of oncocytic cells in nests embedded in a fibrous or myxoid stroma (prototypical morphology of renal oncocytoma). High grade oncocytic tumor of the kidney is also an emerging and recently described entity in kidney neoplasia. The morphology and immunophenotype are similar to oncocytoma. In contrast, both oncocytoma and low grade oncocytic tumour of kidney lack the conspicuous finding of high grade ISUP 3 nuclear features (nucleolar prominence) (He et al. 2018). Renal cell carcinoma of chromophobe type could be ruled out in this case due to absence of CD117 expression and characteristic nuclear irregularities ("raisinoid nuclei"). Hybrid oncocytic chromophobe tumor is currently defined as a variant of chromophobe carcinoma. It may occur in sporadic form or related to oncocytomatosis and Birt-Hogg-Dubé syndrome. CD117 is invariably positive in these tumors and - even if raisinoid nuclei may not be detected (or only focally) -, oncocytic cells may show binucleation and perinuclear halos (Srigley et al. 2013).



**Fig. 2** Low-grade oncocytic tumour of kidney. HE stain shows the transition between a solid oncocytic area with cords of tumor cells in edematous stromal (**a**, 40x; and **b**; 100x). HE stain also shows zones of fresh hemorrhage (**c**, 40x; and **d**, 100x). Stromal edematous areas show frequent hemosiderin-laden macrophages. Immunohistochemistry shows strong and diffuse expression of cytokeratin 7 (**e**, 40x) and absence of c-KIT expression (**f**, 100x). Again, hemosiderin-laden macrophages show brown pigment in edematous stroma

## Conclusion

It is important to recognize additional cases of this emerging entity. The proper use of this new diagnostic category avoids labeling such tumors as unclassified renal cell carcinoma – a broad category with different morphologic features and heterogenous prognosis. At this time, low grade oncocytic tumour of kidney is recognized as an indolent neoplasm with no adverse clinical outcomes reported.

#### Abbreviations

HE: Hematoxylin and eosin stain; ISUP: International Society of Urological Pathology; PAX8: Paired box gene 8

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# Authors' contributions

DAA conceived the idea. DAA was the major contributor to the writing of the manuscript. DAA and ISP diagnosed the case. ISP, CGRAM and MVNX were major contributors for critically revising the manuscript for important intellectual content. The authors read and approved the final manuscript.

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## Availability of data and materials

Not applicable.

## Ethics approval and consent to participate

Not applicable.

#### Consent for publication

Written informed consent was obtained from the patient for participation in the study.

# Competing interests

The authors declare that they have no competing interests.

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