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Oncology

A case report of unilocular cystic mucinous tubular and spindle cell carcinoma with mural tumor nodule

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A B S T R A C T

Mucinous tubular and spindle cell carcinoma (MTSCC) is a rare type of renal cell carcinoma (RCC). Classic type of MTSCC is characterized by small, elongated tubules lined by clear cuboidal or spindle cells with mucinous stroma. The neoplastic cells are always low-grade histological features. But, unclassified variants of MTSCC have also been reported, e.g., mucin-poor, papillary, high grade, and sarcomatoid variants.

We present the first case of simple cyst with mural nodule exhibiting the histological features of mucin-poor MTSCC. We should be aware that MTSCC can arise in a cystic renal lesion.

Introduction

Mucinous tubular and spindle cell carcinoma (MTSCC) is a rare subtype of renal cell carcinoma (RCC). This tumor occurs more frequently in women, with a female-to-male ratio of 3–4:1[1]. Many of MTSCCs are described as low-grade tumor,[1] but case of high-grade histological features and sarcomatoid change may occur. Typical histological pattern of MTSCC is formation of bland tubules, many of which are elongated and merge into cord-like structures, and transitions between tubules and spindle cells are seen.[1] Most of the tumors usually have extracellular mucinous/myxoid stroma, which may be highlighted by Alcian blue stain, but mucin-poor variants of MTSCC have also been reported.[1–3]

Immunohistochemically, MTSCC is consistently positive for PAX2/PAX8 and low molecular weight cytokeratins (CK7, CK18, CK19)[1]. It is also positive for alpha-methylacyl-CoA racemase (AMACR), E-cadherin and vimentin, but often negative for CD10, c-kit, HMB45, Melan A and S100.

According to the past literature, all of the cases of MTSCC show solid cut surface, and to the best of our knowledge, MTSCC with cyst formation has not been reported so far.

Case presentation

A 58-year-old woman was admitted to the Shimane University Hospital for glycemic control. A cyst with mural nodules in the right kidney was detected by contrast computed tomography (CT). The lesion was an unilocular cyst with a maximum diameter of 5 cm and had a solid part that was elevated from the cyst wall. Some foci of microcalcification were found in the cyst wall (Fig. 1a). Retroperitoneal right nephrectomy was performed.

Pathological findings

A cystic lesion of 5.3 × 4.7 × 3.8 cm in size was found in the upper part of the resected kidney. The lesion contained a tan to yellow, solid mural nodule with a maximum diameter of 2.5 cm (Fig. 1b, c, d). The tumor was composed mainly of tightly packed small tubules (Fig. 2a). Slit-like ducts and spindle-shaped cells were also seen in the background of small amount of basophilic mucin (Fig. 2b). There was no significant nuclear atypia and mitosis could not be identified (Fig. 2c). Furthermore, irregular wrinkled nuclei (distinct feature of chromophobe renal cell carcinoma) was not evident (Fig. 2c). Although there was a small highly cellular area of neoplastic cells, but nuclear atypia was not

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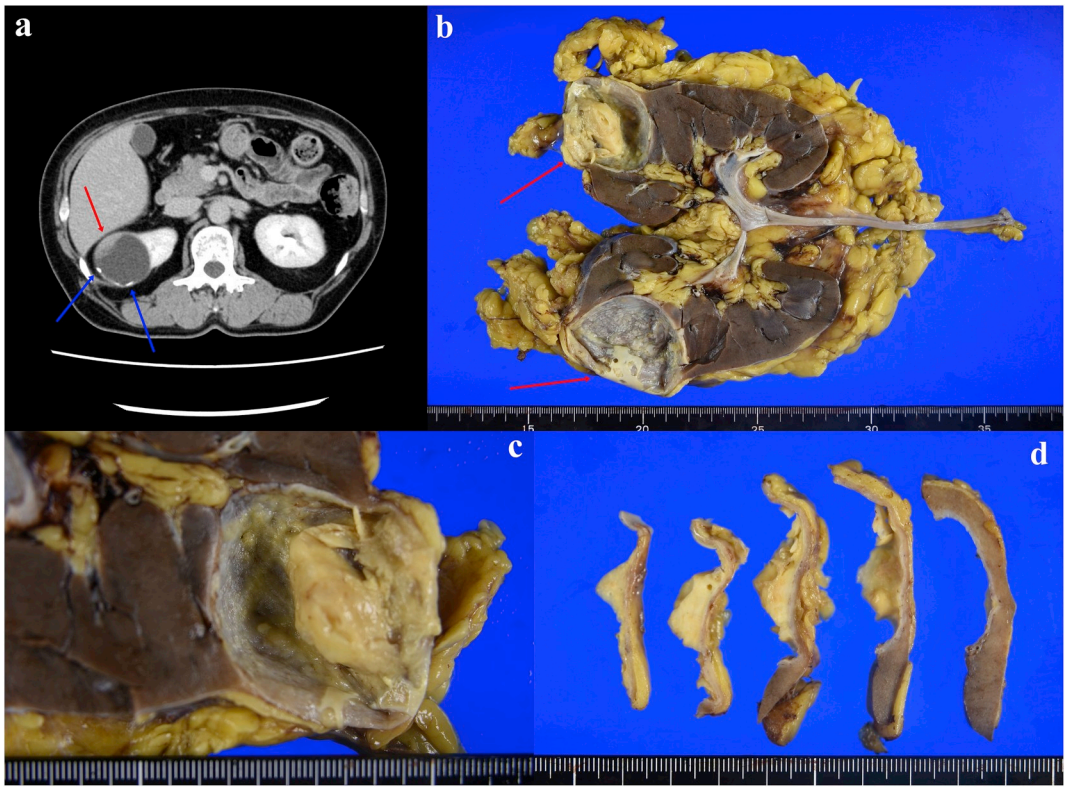

Fig. 1. a: Unilocular cystic tumor of the right kidney. Red arrow: mural nodule of simple cyst, Blue arrow: calcifications of cyst wall. b: Resected kidney. Cystic lesion with mural nodule is seen in the upper part of the right kidney. c: Enlarged view of cystic lesion. d: Cut surface of the cystic tumor. Mural nodule is tan to yellow in color. . (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

significant (Fig. 2d). There was no necrosis in the tumor or invasion beyond the cyst wall (Fig. 2e). A thin area composed of neoplastic cells with a myxoid stroma just beneath the lining epithelium of simple cyst was observed, which was continuous to the mural nodule. The lining epithelium was comprised of low columnar to cuboidal cells which appeared to be benign (Fig. 2f).

Immunohistochemically, neoplastic cells were diffusely and strongly positive for vimentin (Fig. 3a), CK7 (Fig. 3b), E-cadherin (Fig. 3c), PAX8 (Fig. 3d), and AMACR (Fig. 3e). All tumor cells were negative for CD10, c-kit, TFE3, HMB45, Melan A, and S100. The tumor was partially and weakly positive for EMA (Fig. 3f). Considering the histology and the results of immunostaining, we have reached a diagnosis of MTSCC of the mural nodule in the cystic lesion.

Discussion

MTSCC has been accepted as a distinct disease entity since the 3rd edition of World Health Organization Classification of kidney tumors which was published in 2004.[1] MTSCC is now widely known to be positive for AMACR, one of the markers of proximal convoluted tubules.

Clinically, in many cases of this tumor, the patients show good prognosis with histological features of low-grade malignancy.[1] However, aggressive clinical course and/or high-grade morphological findings have been reported.[1] Typical histology of MTSCC is characterized by a mixture of tubular and spindle cell components, and they are separated by variable amounts of mucinous stroma.[1] The tubules are elongated and tightly packed and arranged in parallel.[1]

Mucin-poor variants of MTSCC have also been reported,[1–3] and the tumor of the present case is identical to the mucin-poor variants, but the neoplastic cells did not exhibit high-grade morphological features, and no mitosis were observed. Additionally, the tumor was confined within the cyst wall. This case is expected to have a good prognosis.

Cystic lesions of the kidney are classified into two categories.[4] The first category is characterized by cystic change that usually involves the kidneys in a bilateral and diffuse pattern.[4] There is a risk of RCC to some extent in this category.[4] The second category of cystic lesion is characterized by the presence of an isolated cystic mass not accompanied by any other cystic lesions of the renal parenchyma.[4] Also this category may also develop RCC.[4]

Cystic RCC represents up to 3–14% of all RCCs,[4,5] and can be further classified into multilocular cystic RCC, unilocular cystic RCC, RCC with extensive necrosis, and simple cyst with one or more mural tumor nodules.[4] Cystic RCCs exhibit slow indolent growth and metastasis and recurrence are not common.[5] Cystic RCC characterized by a unilocular cyst with mural nodules is rare, and most of the histological features of the nodules are clear cell RCC, and less frequently papillary RCC.[4,5] To the best of our knowledge, there has not been reported that MTSCC occurred in the mural nodule.

Conclusion

We reported an unusual renal tumor in which the mural nodule in the cyst wall was MTSCC. We comprehensively examined immunostaining, and confirmed the diagnosis of MTSCC. We should be aware that MTSCC can occur as a cystic renal lesion.

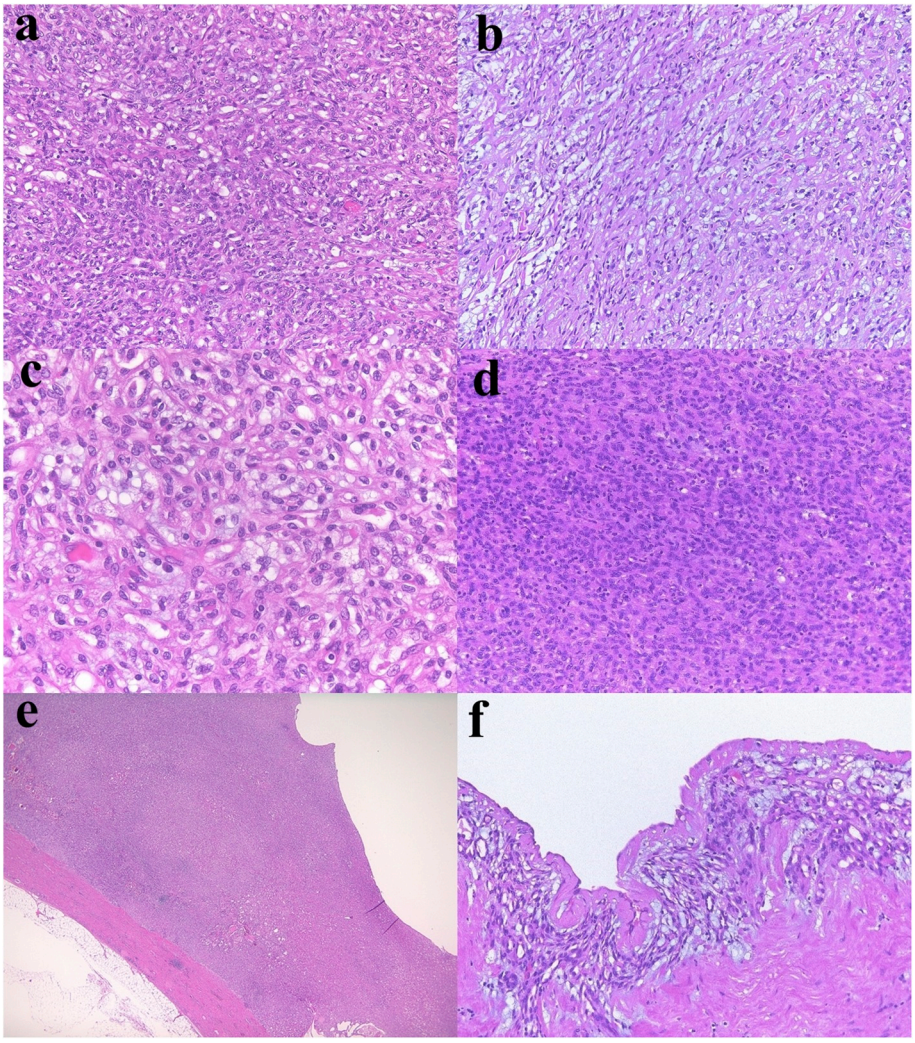

Fig. 2. a: Small tubules are recognized within the tumor. Some tubules are somewhat slit-like structure. b: Area of spindle cell proliferation. Mxyoid stroma can be observed in this area. c: Low grade atypia of neoplastic cells. d: Highly cellular area of neoplastic cells. e. Low magnification of mural nodule. No infiltration beyond the cyst wall by tumor. f: Area of lining cyst. Cyst wall is lined by single layer of columnar cells with scanty cytoplasm, under which tumor cells with myxoid stroma is identified.

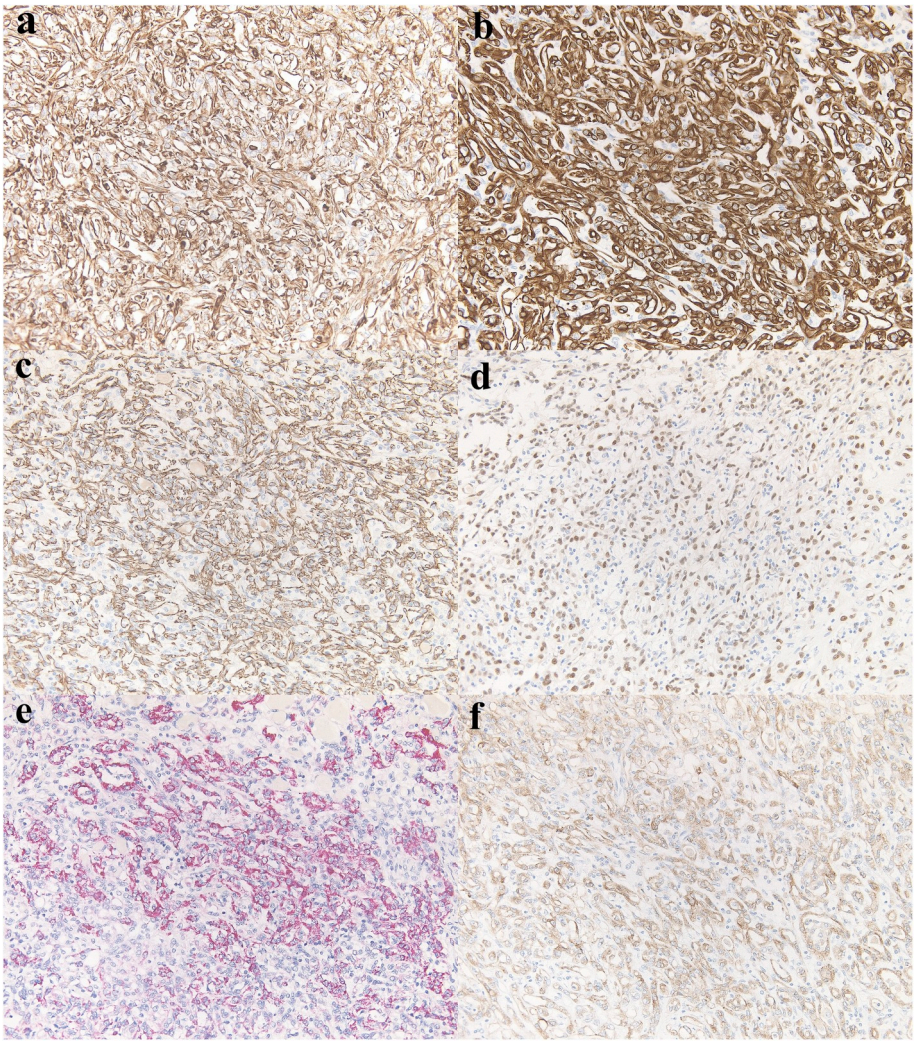

Fig. 3. a: Vimentin, b: CK7, c: E-cadherin, d: PAX8, e: AMACR, f: EMA.

Conflicts of interest

All authors have no perceived conflicts of interest to declare. The authors have no personal financial or institutional interest described in this article.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.eucr.2019.101007.

References

1. Srigley JR, Delahunt B, Eble JN, et al. ISUP Renal Tumor Panel. The International Society of Urological Pathology (ISUP) Vancouver classification of renal neoplasia. *Am J Surg Pathol*. 2013;37:1469–1489.
2. Sokolakis I, Kalogirou C, Frey L, et al. Mucin-poor mucinous tubular and spindle cell carcinoma of the kidney presented with multiple metastases two years after nephrectomy: an atypical behaviour of a rare, indolent tumour. *Case Rep Urol*. 2017, 6597592.
3. Farghaly H. Mucin poor mucinous tubular and spindle cell carcinoma of the kidney, with nonclassic morphologic variant of spindle cell predominance and psammomatous calcification. *Ann Diagn Pathol*. 2012;16:59–62.
4. Truong LD, Choi YJ, Shen SS, Ayala G, Amato R, Krishnan B. Renal cystic neoplasms and renal neoplasms associated with cystic renal diseases: pathogenetic and molecular links. *Adv Anat Pathol*. 2003;10:135–159.
5. Jhaveri K, Gupta P, Elmi A, et al. Cystic renal cell carcinomas: do they grow, metastasize, or recur? *AJR Am J Roentgenol*. 2013;201:292–296.