There is no currently recommended systemic therapy for recurrence or metastasis of RMTSCC. In our investigation, one case was reported with a good response to sunitinib in RMTSCC with low nuclear grade.[16] In contrast, other RMTSCCs with high nuclear grade did not respond to chemotherapy that included sunitinib, axitinib, and temsirolimus. As the second-line therapy, one paper has reported about efficacy of nivolumab in non-clear cell RCC patients including one case of RMTSCC.[17] CT showed a stable tumor at 6 months after nivolumab treatment, but the nivolumab was stopped at 8 months because of tumor progression. In the future, we will report the detail of clinical course after recurrence.

Conclusion

We presented a case of RMTSCC with high nuclear grade. After nephrectomy, metastasis at second lumbar vertebra and lymph nodes recurrence occurred. The prognosis of RMTSCC with high nuclear grade is considered to be unfavorable. Further studies with a large number of cases are required to compile evidence on the follow-up course and therapeutic algorithm in RMTSCC. Additionally, biomarkers to predict the prognosis of RMTSCC are required because recurrence and metastasis occurred even in RMTSCC with low nuclear grade. When diagnosing RMTSCC, clinicians should carefully observe the nuclear grade of the neoplastic cells and the area occupied by these cells with high nuclear grade.

Ethics

We obtained written informed consent from the subject.

Acknowledgement

We thank Mr Shinichi Norimura for his excellent technical assistance.

Conflict of interest

The authors declare no conflict of interest.

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Editorial Comment

Editorial Comment to Case of renal mucinous tubular and spindle cell carcinoma with high nuclear grade

Renal mucinous tubular and spindle cell carcinoma (RMTSCC) is a relatively rare type of malignancy arising from renal parenchyma, which can be characterized by tiny, long tubules lined by cuboidal cells with or without spindled cells separated by pale mucinous stroma.[1] Although

RMTSCC is described in the previous literature as a low-grade tumor, it has a wide range of histological spectrum ranging from low to high grade, sometimes leading to dismal prognosis. To date, number of the literatures are limited up to around hundred, the biological behavior and clinical outcomes, especially metastatic potential of RMTSCC is to be undefined.[1,2] More recently, Wang *et al*. found chromosomal losses and somatic mutations of genes in the Hippo pathway in RMTSCC.[3] They performed an integrative analysis of 907 renal cell carcinoma samples from in-house and TCGA dataset and identified VSTM2A and IRX5 as novel cancer-specific biomarkers in MTSCC.[3] The majority of non-RMTSCC tumors demonstrated negative or low expression of VSTM2A, while IRX5 being suggested as a lineage-specific biomarker, showing moderate to high expression in MTSCC tumors.[3] There are some metastatic RMTSCC cases which are linked to sarcomatoid differentiation.[4,5] Given all these facts, clinicians better be aware that patients with RMTSCC especially with sarcomatoid features should undergo frequent medical checkup according to high grade clear cell carcinoma after surgery even with curative intent.

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DOI: 10.1002/iju5.12089

Conflict of interest

The authors declare no conflict of interest.

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