Front-line pembrolizumab plus axitinib in metastatic Mucinous tubular and spindle cell carcinoma

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Mucinous tubular and spindle cell carcinoma (MTSCC) of the kidney is an uncommon renal neoplasm. Although the prognosis is known to be good, there have been several rare cases of distant metastasis. Due to the rarity of metastatic MTSCCs, standard treatments have not yet been established. We hereby report a case of metastatic MTSCC treated with pembrolizumab plus axitinib as first line chemotherapy. A 59-year-old male patient visited for chest and back pain. Eight months ago, he received left robotic partial nephrectomy with a 3.3cm mass of the left kidney found on an abdomen computed tomography(CT). Histologic examiniation was diagnosed with MTSCC. The tumor was confined to the kidneys with pathologic stage pT1aN0. Immunohistochemical staining for Vimentin, EMA, CD31, D2-40 was positive. Positron emission tomography-CT(PET-CT) showed metastasis in the lung, left supraclavicular and mediastinal lymph nodes, and multiple bone. Biopsy of the left supraclavicular lymph node revealed a recurrence of MTSCC. The patient was started on Pembrolizumab(200mg) and Axitinib(daily 10mg) at 3 week intervals. After the third cycle of chemotherapy, PET-CT showed a stable disease. During chemotherapy, side effects such as hypothyroidism occurred, so synthyroid was administered. After sixth cycle of chemotherapy, PET-CT showed aggressively rapid progression with metastasis on liver, both adrenal gland, spleen with peritoneal carcinomatosis. He received cabozantinib(daily 60mg) as second line chemotherapy, but he showed poor general conditions and platelet count was decreased. Eventually, He died due to multiple organ failure. In conclusion, MTSCC of the kidney is a rare disease with good prognosis, but those with distant metastasis can have a poor outcome. This patient is the first case to be treated with combination of immune check point inhibitor and vascular endothelial growth factor inhibitor in metastatic MTSCC. Stable disease was maintained for about 2-3 months, followed by rapid progression. There is no established treatment for metastatic MTSCC, so more clinical research for treatment is needed.

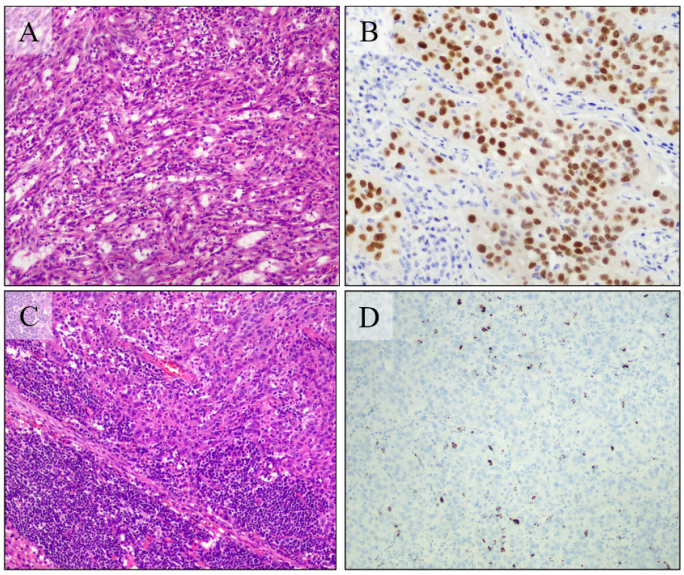

Figure. Histologic features of primary and metastatic tumors. (A) The primary tumor consists of anastomosing tubular structures and spindle cell areas (H&E stain, original magnification: x100). (B) The tumor cells showing diffuse immunoreactivity for PAX-8 (original magnification: x200). (C) The metastatic lesion shows similar growth pattern as primary tumor (H&E stain, original magnification: x100). (D) Immunohistochemical staining for PD-L1 (SP142) showing positivity on the tumor infiltrating inflammatory cells (original magnification: x100).