Case Report

Renal mucinous tubular and spindle cell carcinoma with an aorto-caval mass mimicking metastatic lymphadenopathy

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Abstract

A 54-year-old female had a 9-cm left renal mass with a 12-cm aorto-caval mass lesion mimicking an enlarged lymph node. Retroperitoneal dissection and left radical nephrectomy were performed, and pathology revealed a left renal mucinous tubular and spindle cell carcinoma combined with a retroperitoneal ganglioneuroma. The patient has had no local recurrence or distant metastasis after 3 years’ follow-up. A misdiagnosis of metastatic renal cell carcinoma may be upheld by the primary imaging studies. Even in the targeted therapy era, cytoreductive nephrectomy is still an important step in the diagnosis and treatment of suspicious metastatic renal cell carcinomas.

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1. Introduction

Recent advances in targeted therapy for metastatic renal cell carcinoma (RCC) have produced an expansion of therapeutic options. Neoadjuvant targeted therapies have shown some benefit in advanced renal cell carcinoma and become alternative options before cytoreductive nephrectomy in specific conditions. Here, we report a case of concomitant renal tumor and retroperitoneal mass found at initial presentation. The patient was considered preoperatively to have a metastatic RCC. The pathology report showed a rare RCC entity, mucinous tubular and spindle cell carcinoma (MTSCC) with a retroperitoneal ganglioneuroma, which was different from our prediction.

2. Case report

A 54-year-old female had abdominal fullness and was found to have a left renal tumor 8.5 cm × 8.5 cm × 9 cm in size on computed tomography (CT) images. The tumor was low-density with inner calcification. Additionally, there was another low-density mass lesion located in the inter-aorto-caval area, which mimicked enlarged lymphadenopathy (Fig. 1). No other metastatic lesions were found in the lung, liver, or bones.

First, the aorto-caval mass was dissected retroperitoneally through an abdominal midline incision (Fig. 2). The tumor was a pink, firm, partly encapsulated mass, 12 cm in the longest axis, and could be separated from the aorta and the inferior vena cava. Radical nephrectomy of the left kidney was then performed. The tumor was a well-encapsulated, gray-yellowish myxoid necrotic mass without renal pelvis and renal vessel involvement (Fig. 3).

Microscopically, the retroperitoneal tumor showed some lymphoid tissue, and 17 lymph nodes were counted without...
malignant components. The other part of the mass, which was 9 cm in size, was composed of Schwann cells and ganglion cells. The renal tumor was composed of cuboidal tubular tissue and spindle cells and separated by mucinous stroma (Fig. 4). Staining for vimentin was positive in the tumor, demonstrating its epithelial origin. The α-methylacyl CoA racemase (AMACR) stain showed focal positive areas on the cuboidal tumor cells and implied a renal tubular origin. As the tumor had mucinous components, cytokeratin 7 (CK7) staining was performed, and the positive result excluded a colorectal origin. According to 2004 World Health Organization (WHO) classification of tumors of the urinary system and male genital organs, the renal tumor was classified as an MTSCC.2 The final diagnosis was MTSCC of the left kidney combined with retroperitoneal ganglioneuroma.

The patient received regular outpatient follow-up, and no local recurrence or distant metastasis has been found after 3 years.

3. Discussion

MTSCC was first described in 1998 and was formally included in the 2004 WHO classification.3 MTSCC is predominant in females, generally exhibits low-grade nuclear features, and has a favorable prognosis.3–5 However, some cases with sarcomatoid features may have a poorer prognosis, and adequate sampling of the tumor is required.6,7 MTSCC is mainly diagnosed on histological and morphological grounds, and immunohistochemical studies show mostly specifically positive for RCC marker antigen, vimentin, CK7, and AMACR.6–8
The value of imaging studies for the differential diagnosis of MTSCC is limited. Magnetic resonance imaging reveals signal intensity similar to that of renal parenchyma on T1-weighted imaging, and heterogeneous and slightly high on T2-weighted imaging. MTSCC is also a hypovascular lesion. However, the radiological findings are not specific enough and require differentiation from those of papillary RCC, chromophobe RCC, oncocytoma, and fat-poor angiomylipoma. In this case, CT scanning showed a heterogeneous, low-density tumor with focal calcification, which would undoubtedly be regarded as an RCC.

This case is interesting not only because of the rarity of MTSCC, but also due to the concomitant existence of a renal tumor and retroperitoneal mass. In this era of targeted therapy, the presenting images in this patient might lead to a recommendation of renal biopsy rather than a radical, wide excision of the whole existing tumor. If so, this patient’s diagnosis would have been a metastatic renal tumor instead of a localized T2 tumor. Further, the patient might have had adjuvant therapy for the predicted metastatic tumor. The influence of this misleading diagnosis would have been tremendous.

This case highlights the importance of cytoreductive nephrectomy and possibly metastatectomy. In suitable settings, as complete as possible a removal of the tumor is the best policy when dealing with renal cancer patients.

References