CASE REPORT

Papillary Renal Cell Carcinoma with Extensive Paraaortic Nodal Metastasis Mimicking Malignant Lymphoma

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A 52-year-old woman with abdominal distension underwent computed tomography (CT) that demonstrated extensive paraaortic lymphadenopathy and a right renal mass. Compared to the renal cortex, the lesions exhibited low signal intensity on T1- and T2-weighted images and high intensity on diffusion-weighted magnetic resonance (MR) images. We suspected malignant lymphoma and performed excisional biopsy, which revealed metastatic papillary renal cell carcinoma. Retrospectively, significantly reduced signal on in-phase chemical shift MR images compared to out-of-phase images suggested the presence of intratumoral hemosiderin, a characteristic finding of this entity.

Keywords: chemical-shift MR image, hemosiderin, nodal metastasis, papillary renal cell carcinoma, T2* shortening

Introduction

Papillary renal cell carcinoma (papRCC) is the second most common RCC after clear cell type, has better prognosis than the other cell types,1–7 and has 2 subtypes. Type 1 comprises small basophilic cells that form small papillae and Type 2, relatively large eosinophilic cells that form relatively large papillae. Hereditary lesions are known to be Type 1, and Type 2 lesions are known to have worse prognosis than Type 1 lesions and are typically associated with nodal metastasis.1,6,7 Histologically, papRCC is characterized by hemosiderin within the tumor,4,5 which causes low signal intensity on T2-weighted or echoplanar magnetic resonance (MR) images and apparent signal loss on in-phase images (longer echo time [TE]) of chemical shift images compared to out-of-phase images.8

We present a case of Type 2 papRCC with such prominent nodal metastasis that it was initially misdiagnosed as malignant lymphoma with renal involvement. Retrospective review of MR images revealed apparent signal loss on in-phase chemical shift images that suggested the presence of intratumoral hemosiderin, which could have yielded correct diagnosis at presentation.

Case Report

We examined a 52-year-old woman with a large abdominal mass. Laboratory data on admission was mostly normal, except for slight elevation of serum soluble interleukin-2 (IL2) receptor level (750 U/mL; normal range 160 to 650). No hematuria was evident. Multidetector-row computed tomography (MDCT) (Aquilion 64, Toshiba, Tokyo, Japan) demonstrated extensive retroperitoneal lymphadenopathy at the midabdomen that extended from the level of the esophagogastric junction to above the bifurcation level as well as a right renal mass that appeared to comprise 2 separate components. Left supraclavicular nodal enlargement was also noted. The lesions were relatively homogeneous and showed slight early enhancement and subsequent washout (Fig. 1). The patient underwent MR imaging with a 1.5-tesla clinical unit (Intera Achieva Nova Dual, Philips, Eindhoven, The Netherlands). All lesions showed homogeneous signal intensity resembling that of paraspinal muscle on T1-weighted images. On T2-weighted images with fat saturation, the lesions exhibited higher signal intensity than the paraspinal muscle but slightly lower signal than the normal renal parenchyma. On
Fig. 1. Multidetector-row computed tomography (MDCT) obtained with a total injection of 600 mgI/kg iodine contrast medium in 30 s. GS, gallstone; VC, inferior vena cava; PH, pancreas head. (A) Arterial dominant phase image obtained with 40-s delay. Note extensive retroperitoneal nodal enlargement with slight homogeneous enhancement (arrows) and right renal lesions with partial necrosis (white arrow). The renal lesions consist of 2 separate components, each measuring about 4 cm in diameter. The CT value of the renal mass was 85 Hounsfield units (HU). (B) Equilibrium phase image obtained with 240-s delay. All lesions showed slight washout. The CT value of the renal mass was 70 HU.

diffusion-weighted images (b factor 1000 s/mm²) they showed high signal intensity with calculated apparent diffusion coefficient values of 0.7 to 0.8 × 103 mm²/s (Fig. 2). Based on these imaging findings and slight elevation of the IL2 receptor level, we diagnosed malignant lymphoma involving the retroperitoneal nodes and right kidney. To confirm definitive pathological diagnosis and cell types, we performed excisional biopsy via a small incision for the right renal mass, which revealed relatively large eosinophilic tumor cells that formed large papillae and suggested Type 2 papRCC. Berlin blue staining of the specimen revealed diffuse presence of hemosiderin within the specimen (Fig. 3). Retrospective review of the dual-echo chemical shift MR images showed apparent signal loss on the in-phase (long TE) compared to out-of-phase (short TE) images, suggesting the presence of hemosiderin. The patient was diagnosed with renal cell carcinoma of clinical stage T3aN2M1 and underwent molecular targeted therapy. At the time of writing (18 months after initial presentation), the patient is alive and followed as an outpatient, but tumors show slight progression.

Discussion

Although the prognosis of papRCC is well known to be better than that of clear cell RCC, its Type 2 variant can have extensive nodal metastasis at presentation and a relatively worse prognosis than the Type 1 variant.1-7 Giuliani and colleagues reported a case that initially presented with metastasis of a 2.5-cm papRCC to the cervical lymph node.8 Our case also had extensive nodal metastasis at the retroperitoneum and at the left supravacular region. Retrospectively, low signal intensity on T₂-weighted MR images relative to the normal renal parenchyma and not so prominent vascularity on early-phase MDCT, that is, hypovascularity compared to conventional clear cell RCC, is consistent with the imaging features of papRCC.8-10 However, these findings could also represent malignant lymphoma.11-13 Nevertheless, apparent signal reduction on the in-phase compared to out-of-phase image of chemical shift MR images, representing the presence of intratumoral hemosiderin, is considered specific for papRCC,8 and has not been reported for malignant lymphoma. Dual-echo chemical shift MR imaging has been primarily applied to detect small amounts of fat,14 using significant signal loss on the out-of-phase images as a clue, whereas significant loss on the in-phase images can indicate T₂* shortening and typically suggests the presence of iron or hemosiderin because the echo time of the in-phase image is set longer than that of the out-of-phase image. The somewhat exophytic features of the renal lesion in our case may have been another differentiating point. Such growth is rather rare for malignant lymphoma, with an infiltrative pattern of lymphoma reported within the renal parenchyma without substantial contour deformity.11-13 Meticulous analysis of all MR imaging findings, particularly chemical shift images, could have led us to a correct diagnosis in our case.

In conclusion, radiologists need to recognize that the Type 2 variant of papRCC can present with extensive nodal metastasis that mimics malignant
Fig. 2. Magnetic resonance (MR) images obtained with 4-channel phased-array coil. GS, gallstone; VC, inferior vena cava; PH, pancreas head. (A) Fast spin-echo T2-weighted image repetition time [TR]/echo time [TE]/echo train = 1073 ms/70 ms/24) obtained with fat suppression and respiratory triggering. All lesions show higher signal intensity than the paraspinal muscle but slightly lower signal than the left renal parenchyma. The GS is out of slice and not visualized. (B) Gradient-echo T1-weighted in-phase chemical shift image (TR/TE/flip angle [FA] = 164/4.2/75°). Note punctuate low signal areas in the nodal and renal lesions (arrows). VC is collapsed due to breath-holding. (C) Gradient-echo T1-weighted out-of-phase chemical shift image (TR/TE/FA = 164/2.3/75°). The low signal spots seen in 2B are not evident, and some spots of high signal intensity are seen instead (arrows). (D) Subtraction image (= 2B−2C). Note areas of low signal intensity that suggest focal signal loss on in-phase image (arrows). (E) Echoplanar diffusion-weighted image (TR/TE = 1500/72) with b-factor of 1000 s/mm². All nodal and renal lesions show high signal intensity. Calculated apparent diffusion coefficient was approximately 0.7 × 10⁻³ mm²/s.

lymphoma, and apparent signal reduction on the in-phase image of chemical shift MR images, which suggests the presence of hemosiderin, is a strong clue to papRCC. Careful interpretation of chemical shift MR images may be useful in differentiating these 2 entities.

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Fig. 3. Berlin blue staining of the pathological specimen (original magnification × 200). Black or dark blue dots indicate hemosiderin granules located in both tumor cells and macrophages (foamy cells).

References