INTRODUCTION

Malakoplakia is a rare chronic inflammatory condition that results from defective phagolysosomal activity. Malakoplakia usually affects the urinary tract, and immunosuppression is a predisposing factor in most patients. A 78-year-old woman undergoing long-term steroid treatment presented with right flank pain. CT demonstrated a large, multilocular cystic mass with focal enhancing solid portion in the right kidney and retroperitoneum. The patient underwent ultrasonography-guided biopsy for enhancing the solid portion, and pathologic examination revealed malakoplakia.

CASE REPORT

A 78-year-old woman was hospitalized in our institution because of right flank pain aggravated by moving for 5 days before admission. She had a history of long-standing rheumatoid arthritis and had been taking prednisolone, 5 mg daily for 12 years. On physical examination, right side abdominal tenderness was elicited. Laboratory tests revealed blood urea nitrogen...
of 34.2 mg/dL, serum creatinine of 0.97 mg/dL, white blood cell (WBC) count of 27400/μL, erythrocyte sedimentation rate of 31 mm/hr and C-reactive protein 157.16 mg/L, suggesting an acute infectious process. Urine analysis revealed pyuria (30–60 WBC/high power field) and urine culture was positive for *Escherichia coli*.

CT examination showed a large, multilocular cystic mass in right kidney and perirenal space with irregular and thick wall and septa, extending into the right retroperitoneum, bare area and right psoas muscle. Focal enhancing solid portion was demonstrated at the cystic mass in the right kidney (Fig. 1A, B). There was no abnormality along the upper urinary collecting system and the bladder. The complicated cystic lesion was initially considered most likely as an abscess involving right kidney and retroperitoneum. So, ultrasonography (US)-guided aspiration was performed to confirm the presence of turbid gray-col-

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**Fig. 1.** Renal malakoplakia with wide retroperitoneal extension in a 78-year-old woman. Axial (A) and coronal (B) contrast enhanced CT scan shows a large multilocular cystic mass with irregular wall and septa (arrowheads) in right kidney and perirenal space. The mass extends into the right retroperitoneum, bare area and right psoas muscle. Focal enhancing solid portion is demonstrated in the right kidney (arrow). On follow-up CT scan (C) obtained after 4 months with antibiotic treatment, the complicated cystic lesion is markedly reduced. And the enhancing solid portion is also decreased in size (curved arrow).

**Fig. 2.** Photomicrographs of malakoplakia.

**A.** There are many scattered histiocytes (von Hansemann cells) containing intracytoplasmic lamellated basophilic inclusions, called Michaelis-Gutmann bodies (black arrows) (× 1000).

**B.** A von Kossa stain highlights numerous Michaelis-Gutmann bodies (× 200).
Renal Malakoplakia with Wide Extension into the Retroperitoneum

Malakoplakia is a rare chronic granulomatous benign disease. It may affect any organ of the body but primarily affects the genitourinary tract (1). Other locations include the retroperitoneum, gastrointestinal tract, central nervous system, female genital tract, lung, pleura, pancreas, spleen, lymph node, adrenal gland, and vertebra (1-3). Most patients have positive urine cultures, usually with Gram-negative bacteria and a predisposition to immunosuppression such as solid organ transplantation, autoimmune disease requiring long-term steroid use, chemotherapy, malignancy, alcohol abuse and diabetes mellitus (1, 3).

Malakoplakia results from defective phagolysosomal activity with incomplete bacteria digestion, mostly *Escherichia coli* and *Klebsiella*, by macrophages and monocytes. Bacterial debris accumulates in the cytoplasm of these cells and usually becomes mineralized. This leads to basophilic inclusion structures with surrounding clear halos i.e., Michaelis-Gutmann bodies, which are pathognomonic for diagnosis (1, 3, 5).

There is a peak incidence in the fifth to seventh decades at presentation and a female predominance with a female to male ratio of 4:1 (1, 2). Even though the symptoms may vary according to the affected organ, malakoplakia is commonly associated with urinary tract infections, including acute renal failure in renal malakoplakia (3).

In most patients with malakoplakia, lesions are initially identified at imaging studies in various clinical settings, including an abdominal mass, hematuria, renal failure, bladder irritability or persistent urinary tract infection despite appropriate antibiotic therapy (1-5). Imaging features of malakoplakia are nonspecific and variable. Malakoplakia can present as a diffuse infiltrative disease, sharply demarcated solitary mass, or ill-defined solid masses. The lesion demonstrates variable echogenicity at US, and mildly enhanced, heterogeneous attenuation at CT. In the case of renal parenchymal malakoplakia, imaging studies commonly demonstrate the enlarged, irregularly contoured kidneys and dedifferentiation of corticomedullary junction. Decreased excretion of contrast is more pronounced in cases with extensive parenchymal involvement. Parenchymal calcification is rare (1, 3, 7). Perinephric extension and renal vein thrombosis have been reported (1). Magnetic resonance imaging features include poorly defined multiple low signal intensity nodules on all sequences with intervening fibrous stroma (3, 8). Focal renal lesions are often misdiagnosed as a malignancy such as necrotic renal cell carcinoma. In imaging studies for renal malakoplakia, differential diagnosis includes xanthogranulomatous pyelonephritis, local abscess, granuloma, lymphoma and primary or metastatic tumors (3, 6).

Malakoplakia rarely extends to other organs. However, the presence of malakoplakia in retroperitoneal space is almost always due to direct extension from an adjacent organ, mainly urinary tract malakoplakia (2). Hence, we could characterize our patient as having renal malakoplakia with wide retroperitoneal extension.

There is no established treatment for malakoplakia. However, there are treatment options including antibiotics, surgical excision or a combination of both (2). Successful medical management has been reported with antimicrobial agents (4, 5). Malakoplakia is a highly treatable disease when detected in its early stage. Immunosuppressive drugs should be discontinued as far as possible (3). Because renal malakoplakia may mimic renal cell carcinoma on imaging studies, a percutaneous or an open biopsy helps to diagnose a treatable disease and prevent unnecessary surgery.

In conclusion, renal malakoplakia should be considered in the differential diagnosis in patients with urinary tract infection, fever, abdominal pain, and renal mass like lesion, especially in patients that have a risk factor of immunosuppression. As shown in this case, renal malakoplakia can present as extensive retro-
peritoneal mass and often be confused with malignancy.

REFERENCES