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Malakoplakia of the Ureter and Bladder

Bruce R. Baumgartner and Ravi Alagappian
Department of Radiology, Emory University School of Medicine, Atlanta, Georgia, USA

Abstract. Malakoplakia is an uncommon granulomatous inflammatory disease that most often involves the urinary tract. Typically, there is an associated urinary tract infection (UTI) by coliform organisms. Histologically, the Michaelis-Gutmann bodies are the hallmark of this disease. Radiographically, malakoplakia may simulate other inflammatory processes or even neoplasm as demonstrated in these two cases.

Key words: Malakoplakia — Urinary tract — Bladder.

Malakoplakia, literally meaning "soft plaque," is an uncommon granulomatous disease first described in 1902 by Michaelis and Gutmann. It is characterized by aggregations of histiocytes containing pathognomonic, intracellular calcifications known as Michaelis-Gutmann inclusion bodies [1]. Malakoplakia has been described in practically every organ system of the body, but most commonly affects the urinary tract, and, in particular, the bladder [1]. Two cases of malakoplakia involving the urinary tract are described along with their associated radiographic findings.

Case Reports

Case 1

An 86-year-old woman, with a history of right upper lobe resection 3 years earlier for small cell carcinoma, presented with gross hematuria of 3 months' duration. The patient denied any increased urinary frequency, urgency, or dysuria and had no previous history of urinary tract disease. Biopsy of a retrovesical mass at cystoscopy revealed normal bladder mucosa, but showed submucosal, cellular infiltration suggesting a diagnosis of metastatic small cell carcinoma to the bladder. The patient was referred here for further evaluation of her bladder mass.

Admission physical exam revealed a firm, rounded, midline suprapubic mass extending to the umbilicus. Urinalysis and urine culture indicated an *E. coli* urinary tract infection (UTI). Intravenous urogram (IVU) revealed compression on the bladder dome (Fig. 1A). A computed tomographic (CT) scan demonstrated a 6–7 cm retrovesical mass extending along the posterior superior aspect of the bladder and the anterior portion of the uterus (Fig. 1B). There was no evidence of ureteral obstruction or adenopathy. Cystoscopy was performed revealing a centrally ulcerated mass in the posterior aspect of the dome of the bladder which was biopsied; a transvaginal needle biopsy of the retrovesical mass was also performed. Both specimens were diagnostic for malakoplakia, showing a mixed inflammatory infiltrate composed of lymphocytes, histiocytes, plasma cells, and Michaelis-Gutmann inclusion bodies demonstrated by periodic acid-Schiff's stain. The patient was discharged on antibiotics for her UTI but never returned to this hospital for follow-up.

Case 2

A 64-year-old man, with a history of basal cell nevus syndrome, presented with a 14-month history of progressive decreased force of urinary stream and increased hesitancy. He denied any symptoms of flank pain, fever, or hematuria. IVU revealed a moderate postvoid residual and questionable partial obstruction of the right ureter. A diagnosis of benign prostatic hypertrophy was made, and the patient was admitted for a transurethral prostatectomy and retrograde pyelogram (RPG). There was no evidence of UTI at this time. RPG demonstrated a right hydroureter with caliectasis and three filling defects in the distal two thirds of the right ureter (Fig. 2). The findings were felt to be suggestive of either a polypoid tumor of the ureter or chronic inflammatory process. Urine cytology from the right ureter was negative for malignancy. Subsequent admissions for similar complaints were accompanied by documented *E. coli* urinary infection. The patient eventually underwent segmental resection of his right ureter and ureteroureteral anastomosis. Pathology of the lesions in the resected section revealed multifocal malakoplakia associated with ureteritis. The

Address offprint requests to: Bruce R. Baumgartner, M.D., Department of Radiology, Emory University School of Medicine, 1364 Clifton Road, NE, Atlanta, GA 30322, USA

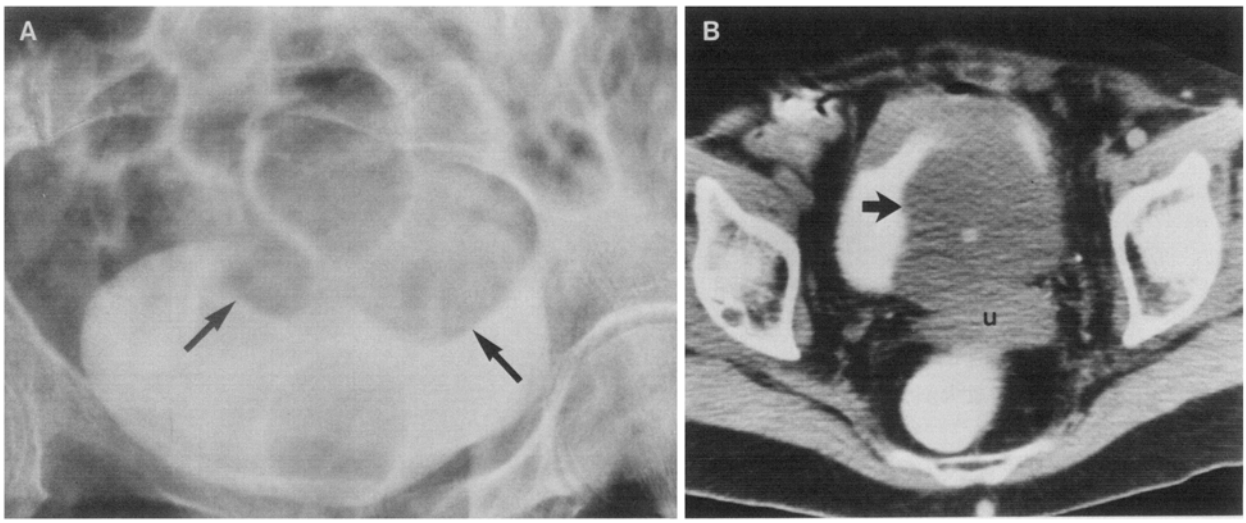

Fig. 1. **A** Malakoplakia: AP view of the bladder during IVU demonstrates a lobulated mass (*arrows*) compressing the dome of the bladder (Foley catheter is in place). **B** Axial CT image reveals the soft tissue mass (*arrow*) that involves the posterior–superior aspect of the bladder and is contiguous with the uterus (*u*).

patient did well postoperatively and was discharged without further complications.

Discussion

There have been approximately 200 cases of malakoplakia reported, since the entity was first described in 1902 by Michaelis and Gutmann. Although this entity has been described in nearly every organ system of the body, the vast majority of cases involve the urinary tract. Stanton and Maxted, in their review of 153 cases of malakoplakia, found 40% of cases to involve the bladder, 16% the renal parenchyma, 11% the ureter, 10% the renal pelvis, 3% the ureteropelvic junction, and 2% the urethra [1–3]. Malakoplakia of the urinary tract affects women four times more frequently than men, while the incidence of disease in other areas of the body occurs more frequently in men [1]. The peak age of incidence is in the fifth decade for women and in the seventh decade for men [1, 4].

Malakoplakia is a granulomatous inflammatory disease. Macroscopically, the yellow-brown lesions may have a varied appearance. Descriptions of bladder malakoplakia include flat, round plaques, nodules, papillary lesions, masses with necrotic calcified centers, as well as trabeculated, polypoid, tumor-like, and hemorrhagic masses [4]. The size of the lesions may vary from a few millimeters to several centimeters [5]. Microscopically, the lesion is characterized by submucosal aggregations of histiocytes with granular cytoplasm (von Hansenmann cells), often mixed with acute inflammatory cells (i.e. lymphocytes, neutrophils, and plasma cells) [4]. Within the cytoplasm of the von Hansenmann cells, intracellular inclusion (Michaelis-Gutmann) bodies and numerous phagolysosomes may be found [4]. Although there may be no Michaelis-Gutmann bodies in the early (prediagnostic) histologic phase of malakoplakia and very few Michaelis-Gutmann bodies in the late (fibrosing) phase, they were considered the hallmark of the lesion and their presence is generally considered necessary in making the diagnosis of malakoplakia [4, 6, 7].

The etiology of malakoplakia remains uncertain, although many theories regarding its pathogenesis exit. Most case of urinary tract malakoplakia initially present as UTIs. The association between malakoplakia of the urinary tract and infection by coliform organisms is well-described. Several animal models involving injection of Boivin antigen extract of *E. coli* cell wall into kidneys of rats have reproduced malakoplakic lesions with Michaelis-Gutmann bodies and suggest a relationship between renal parenchymal malakoplakia, xanthogranulomatous pyelonephritis, and megalocytic interstitial pyelonephritis [4]. Infection with coliform organisms, however, does not seem to entirely account for the development of malakoplakia. Malakoplakic lesions may be found in the urinary tract, as well as in other sites, apparently unrelated to such infections. The incidence of urinary tract malakoplakia is disproportionately low when compared to the frequency of UTIs, and the organisms associated with malakoplakic lesions do not appear to show increased pathogenicity to account for this discrep-

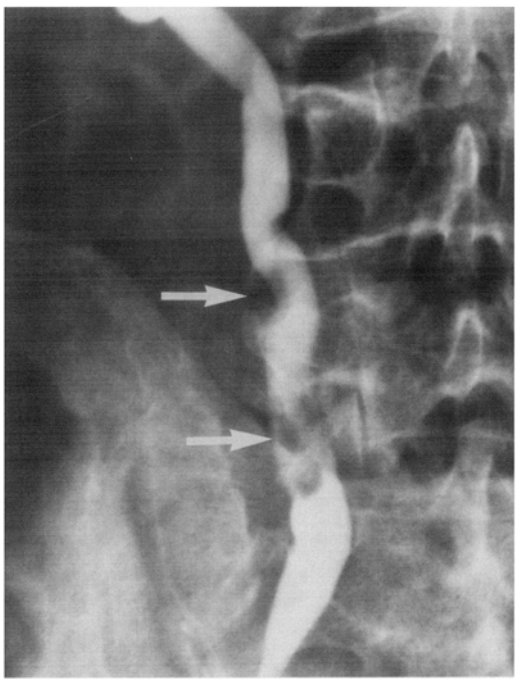

Fig. 2. Malakoplakia: RPG demonstrates deformity of the ureter with several polypoid filling defects (*arrows*).

ancy [8]. There is a strong correlation between the development of malakoplakia and the altered host-immune response. Forty percent of the 153 cases reviewed by Stanton and Maxted had either an intercurrent systemic illness, carcinoma, immune deficiency syndrome, or autoimmune disease [1]. An altered host-macrophage response to infection by microorganisms resulting in impaired digestion within phagolysosomes has also been hypothesized [4].

These two cases demonstrate how difficult the radiological diagnosis of malakoplakia can be. CT findings in the first case demonstrated a mass between the bladder and the uterus. The differential diagnosis for this lesion, in addition to metastatic lung carcinoma, would include primary bladder or uterine neoplasm. RPG findings in the second case demonstrating partial right ureteral obstruction with multiple filling defects are more suggestive of a neoplastic process than a multifocal inflammatory process. In both patients, *E. coli* UTIs and radiological findings revealing urinary tract masses should have suggested the possibility of malakoplakia. While these findings are by no means pathognomonic for malakoplakia, in the clinical setting of recurrent urinary infections, that diagnosis should be considered. CT has also been shown to be useful in determining extent of disease and monitoring response to conservative therapy [5].

The first case also illustrates that histologic findings, although essential for diagnosis, may not always be conclusive. The tissue diagnosis of malakoplakia may require special stains and is dependent on the acumen and experience of the individual pathologist.

It is important to consider and recognize malakoplakia as it may often be treated medically with success, thus avoiding unnecessary surgery. In cases when obstruction has occurred, however, surgical intervention is necessary, despite an accurate diagnosis. These two cases serve to illustrate both of these points.

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