

방광에 발생한 Malakoplakia

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Malakoplakia in the Urinary Bladder

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Malakoplakia is a rare chronic granulomatous disease, which was originally described in the urinary bladder, but can involve many other organs and soft tissues. Malakoplakia is often associated with immunosuppression or immunodeficiency and is believed to be caused by an alternation in the bacterial phagocytic system. Histologically, the presence of Michaelis-Gutmann bodies is pathognomonic. We report on a case of malakoplakia of the bladder in a 62-year-old female.

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Malakoplakia was first described by Michaelis and Gutmann in 1902 and subsequently named by von Hansemann in 1903.^{1,2} The term makakoplakia is derived from the Greek words “malakos”, meaning soft, and “plakos”, meaning plaque.² It is most commonly seen in the urinary bladder (40%), although it has been reported in other locations, including the renal parenchyma, ureter, testis, prostate, epididymis, gastrointestinal tract, retroperitoneum, bone, adrenal gland, brain, and liver.³ It is often misdiagnosed as a malignant condition. We present a 62-year-old woman who was diagnosed with malakoplakia.

CASE REPORT

A 62-year-old woman was referred to our institution from another hospital because of recurrent urinary tract infection. She had history of mild hypertension and diabetes mellitus

for 10 years. She has no history of immunosuppression or surgery.

She suffered from dysuria, frequency, and urgency in the last 30 days. There was no fever or chills during this period and her physical examination was normal. The results of routine blood tests were unremarkable. Urinalysis revealed pyuria and microscopic hematuria. However, urine culture showed no growths. No abnormality was detected in radiologic studies of the upper urinary tracts. Cystourethroscopy revealed multiple, soft, yellow plaques on the entire wall of the bladder (Fig. 1). Multiple biopsies were taken. Microscopic examination showed mixed inflammatory cell infiltrate with histocytes containing the characteristic Michaelis-Gutmann bodies of malakoplakia (Fig. 2). Conservative treatment with fluoroquinolones, bethanechol, and vitamin C was initiated and continued for a 3-month period. All her symptoms including dysuria,

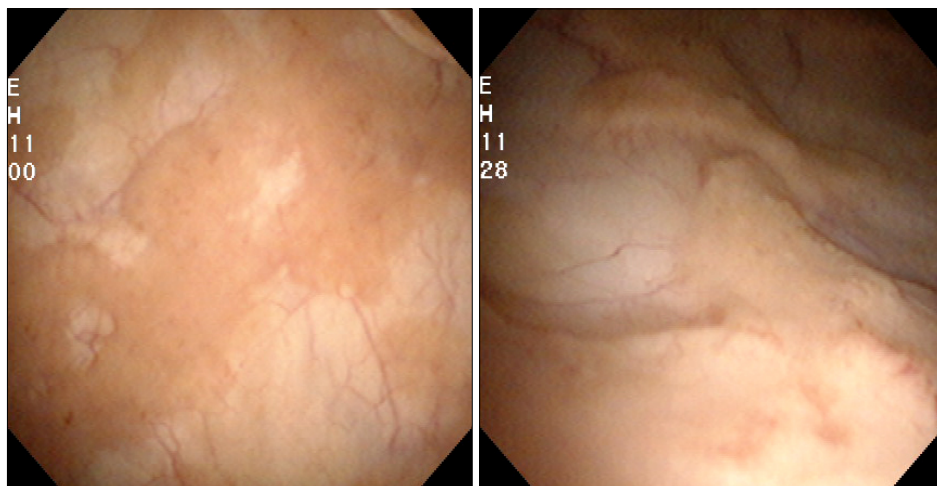


Fig. 1. Cystourethroscopy revealed multiple, soft, yellow plaques on the entire wall of the bladder.

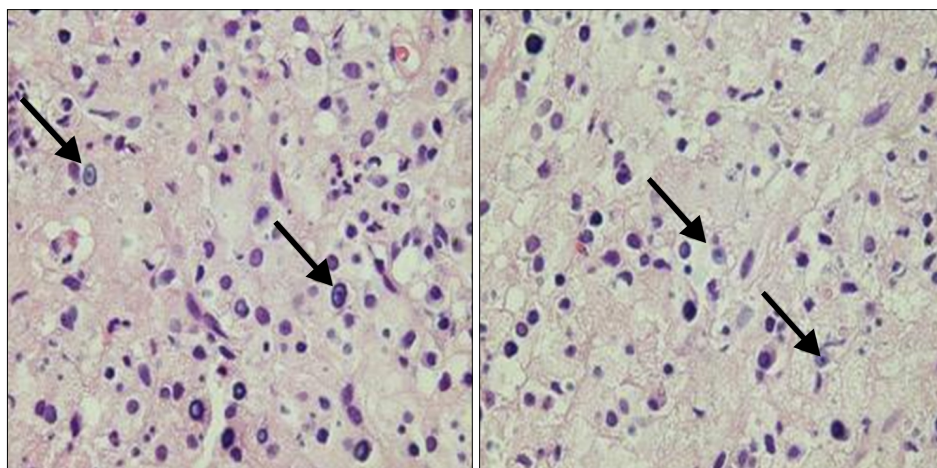


Fig. 2. Michaelis-Gutmann body (arrows) ($\times 400$).

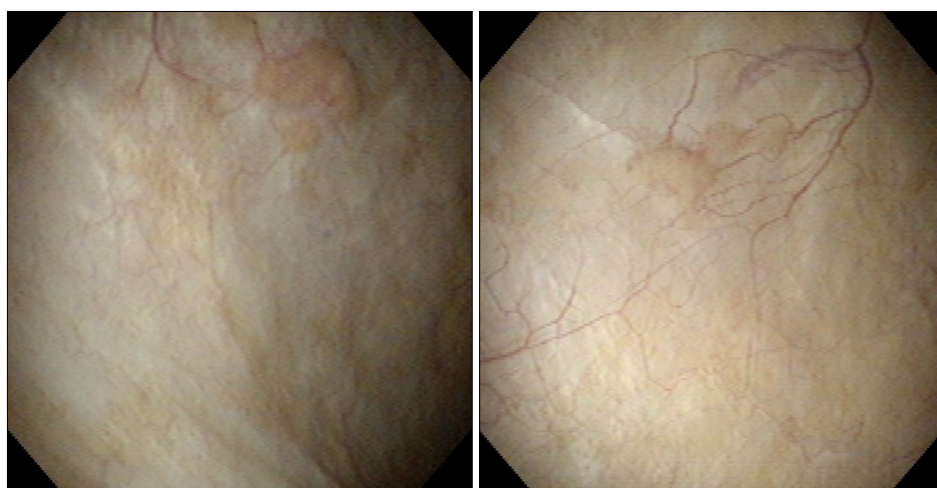


Fig. 3. Cystourethroscopy still revealed the presence of yellow mucosal lesions but their number and size was decreased.

frequency, and urgency disappeared and her urinalysis was normal after 3-months. A follow-up cystourethroscopy still revealed the presence of yellow mucosal lesions but their

number and size was decreased (Fig. 3). Resection of the bladder lesions was performed. In a follow up 6-months later, no lesions were seen on a cystourethroscopy (Fig. 4).

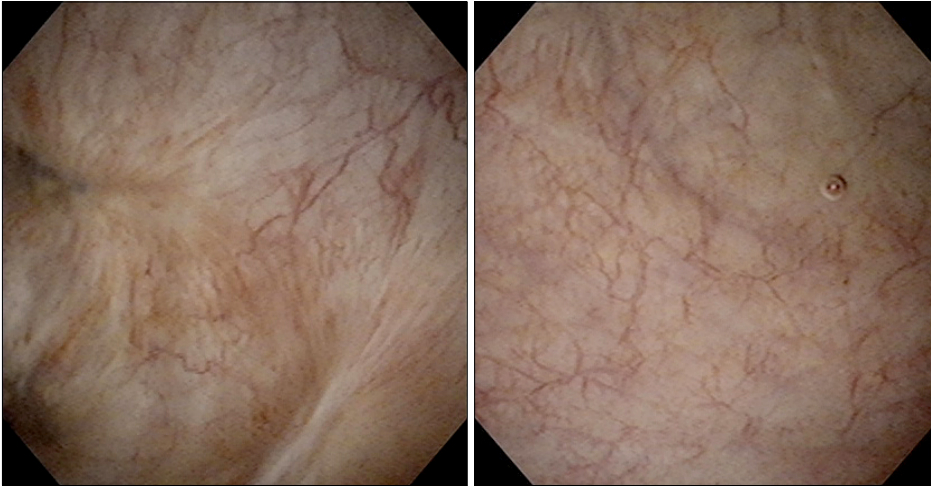


Fig. 4. No lesions were seen on a cystourethroscopy.

DISCUSSION

Malakoplakia is rare inflammatory disorder seen mostly in the genitourinary tract (58%), especially the bladder. It has also been described in the colon, liver, lungs, bones, brain, conjunctiva, pancreas, skin, tonsils, and adrenal gland. It is more common in females, with a female to male ratio of 4:1.³

It is more commonly found in the immunodeficient population such as those with HIV/AIDS, those with autoimmune disease or post-transplant patients.^{4,5}

The majority of patients with malakoplakia present with histories of chronic urinary tract infections, commonly with *Escherichia coli*. Other bacteria as well as mycobacteria and fungi can also cause malakoplakia.

It is believed that malakoplakia represents an unusual granulomatous response to bacterial infection, in which macrophages fail to phagocytose bacteria properly.

Histologically, it is featured by sheets of histiocytes characterized by small nuclei without prominent nucleoli and foamy cytoplasm. In the cytoplasm of some of these cells, round concretions are observed that are highlighted by periodic acid-Schiff histochemical staining. These structures are called the Michaelis-Gutmann bodies, which are pathognomonic.^{6,7}

The symptoms and sign of malakoplakia vary widely, depending on the organ system involved. Bladder malakoplakia most commonly presents similar to bladder tumor or acute cystitis. Renal parenchymal malakoplakia has been seen to present with fever, flank pain, and/or a palpable flank mass. Testis malakoplakia most commonly

presents similar to acute epididymitis, orchitis or epididymo-orchitis.

Treatment of bladder malakoplakia can be treated medically. Medical treatment is based on the eradication of *E. coli* infections that are believed to cause the malakoplakia manifestations. The best antibiotics are those that achieve high intracellular levels such as fluoroquinolones and trimethoprim-sulfamethoxazole.^{8,9} In addition to these agents, bethanechol and vitamin C are believed to enhance phagocytic bactericidal activity by increasing cyclic guanosine monophosphate levels.³ Persistence of malakoplakia may require resection. However, as malakoplakia has a tendency to persist or recur, long-term follow up is needed.

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