



# Isolated ureteral malakoplakia: A conservative endoscopic approach with a strict follow-up

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## ABSTRACT

Ureteral malakoplakia is a rare pathological entity. We report the case of a 54-year-old woman with a single ureteral malakoplakic lesion. Patient presented with history of recurrent urinary tract infections and asymptomatic dilatation of right pelvis. Radiological investigations showed a right lower ureteric filling defect without bladder or kidney involvement. A first uretero-renaloscopy allowed an extirpative biopsy, with a histopathologic diagnosis of malakoplakia. Second-look uretero-renaloscopy showed only a minute area of hyperemic mucosa that was biopsied and coagulated, showing a residual focus of malakoplakia. At 12-months, imaging and blood test demonstrated reduction of hydronephrosis, serum creatinine recovery and no recurrences.

## 1. Introduction

Malakoplakia is a rare granulomatous disease that can occur in the urinary tract, however the involvement of nearly every organ system has been reported.<sup>1</sup> It is described as a self-limiting chronic granulomatous disease which usually carries a good prognosis.<sup>1,2</sup> In the genitourinary tract, it mainly affects the bladder, and less commonly the kidneys and ureters.<sup>3,4</sup> We present a case of a single ureteral malakoplakic lesion in a 54-year-old woman, without bladder involvement.

## 2. Case presentation

Patient presented with a history of recurrent urinary tract infections, one of which complicated with urosepsis, and a left ureterocele. She had a smoking history of 10 cigarettes/day for 20 years. Her remaining past medical history was unremarkable. Ultrasonography confirmed a left ureterocele and detected an asymptomatic right kidney hydronephrosis. CT urography showed a grade 2 hydronephrosis of the right kidney, supported by a 12-mm solid tissue, 25-mm from the ureteral orifice (Fig. 1). Cystoscopy confirmed the presence of a left ureterocele but no other significant findings. Blood tests highlighted elevated serum creatinine (1.53 mg/dL). Urine analyses were normal, and urine culture

negative. Urine cytology was also negative. Right retrograde ureteropyelography, performed after a 3 month delay due to a SARS-CoV-2 infection, showed a 3-cm filling defect along the pelvic tract of the ureter, with associated ipsilateral hydronephrosis (Fig. 2). With the uretero-renaloscope, it was possible to reach the smooth, bulky gray-white lumen-infiltrating lesion that showed a small base of implantation. No other lesions were present along the ureter. Wash fluids and multiple biopsies were taken for cytologic and pathological analysis, respectively. Subsequent laser ablation of the residual lesion and coagulation of the base of implantation were performed. A mono-J catheter was placed in the right ureter and removed prior to discharge. No malignant tumour cells were found at cytological analysis. Immunohistochemical investigations (AE1/AE3, CAM5.2 and Kp1) were performed on the pathological pieces revealing a diffuse infiltrate of Kp1 positive histiocytes with eosinophilic and granular cytoplasm (Von Hansemann histiocytes) with intracytoplasmic inclusions of spherical aspect with concentric laminae, PAS and Von Kossa positive stain (Michaelis-Gutmann bodies). The urothelial lining was free of atypia. The final histopathological diagnosis was malakoplakia (Fig. 3). During the first control, after 2 months, the right renal ectasia had resolved, and the creatinine levels returned within normal limits. Furthermore, a second look uretero-renaloscopy showed at the level of previous procedure only a

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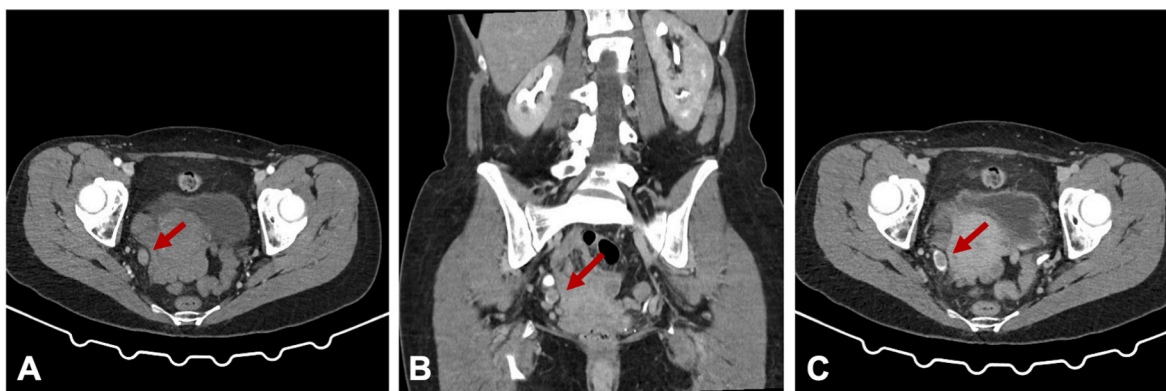


Fig. 1. CT urography that showed a 12-mm solid tissue, 25-mm away from the right ureteral papilla. A: Arterial Phase. B: Coronal Plane. C: Late Phase.

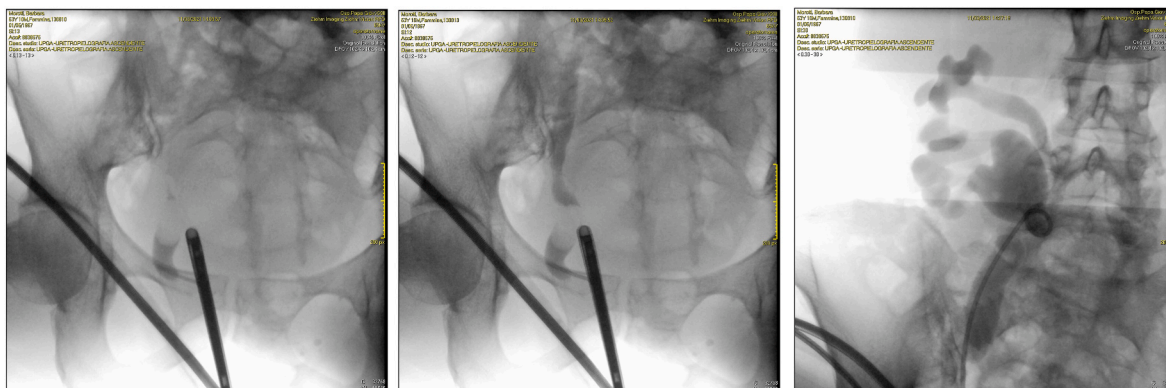


Fig. 2. Right retrograde ureteropyelography showed a 3 cm long filling defect in the pelvic tract of the ureter, with associated ipsilateral hydronephrosis.

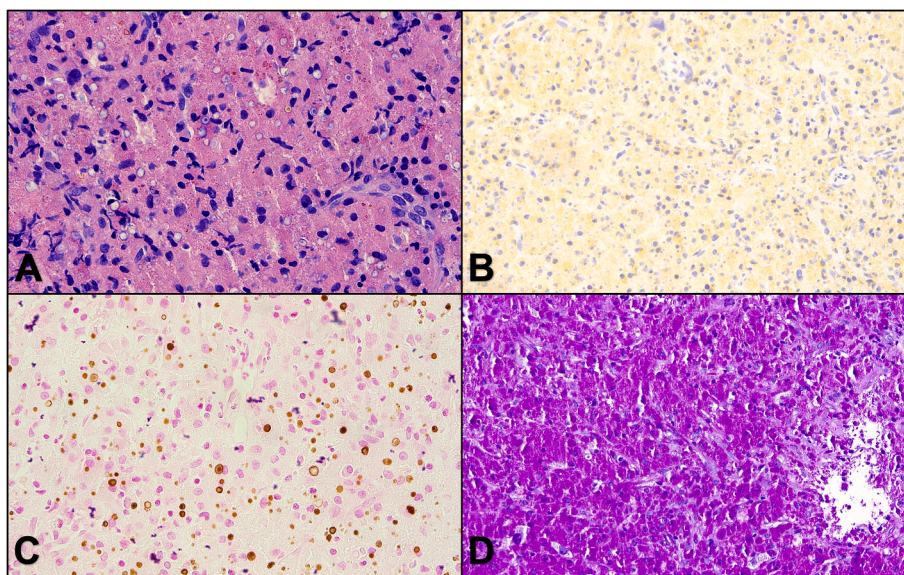


Fig. 3. A: accumulation of histiocytes with granular eosinophilic cytoplasm (von Hanseman cells), 40x; B: immunostain for histiocytes (Kp1 - CD68); C: the von Kossa stain for calcium highlights Michaelis-Gutmann bodies in brown, characteristic intracytoplasmic inclusion, 40x; D: Hanseman histiocytes react with the periodic acid-Schiff stain. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

millimetric area of hyperemic mucosa that was biopsied and coagulated. The anatomopathological analysis were once again compatible with malakoplakia. We established a strict follow-up with upper urinary tract imaging every 6 months. After a 12 month-follow up, the patient presented no recurrences, a normal renal function and no hydronephrosis at

ultrasonography.

### 3. Discussion

The exact etiology of malakoplakia is unknown however *Escherichia*

*coli*'s infection is the most common condition associated with malakoplakia.<sup>1</sup> The most frequent presentation of malakoplakia is irritable bladder symptoms. Our patient reported a history of recurrent urinary tract infections, one of which complicated with urosepsis.

Malakoplakia is more frequent in the bladder than in the renal pelvis or ureter. Upper urinary tract lesions when present, are usually associated with bladder involvement.<sup>1,3,5</sup> When malakoplakia affects the bladder, it can be easily diagnosed with a cystoscopy. Usually, lesions are achromatic, gray-white with sharp edges rising on the mucosa, and cystoscopy-guided biopsy is diagnostic.<sup>1</sup> After an initial diagnosis of bladder malakoplakia, as this disease may affect the whole urothelial mucosa, a CT scan should be performed in order to investigate also the upper urinary tract.

Here we present a case of a single ureteral malakoplakic lesion, resulting in hydronephrosis, and not associated with bladder lesions at diagnosis. In the absence of bladder lesions, such a diagnosis was clearly unexpected. Uretero-rensoscopy with biopsy allowed to distinguish between malakoplakia and urothelial malignancy. This case study further highlights the need for a first step endoscopic evaluation of the upper tract, as imaging and cytology are not always sufficient for the diagnosis or the risk stratification of the disease. Indeed, the endoscopic aspect of the lesion, presenting as a smooth, bulky lesion albeit characterized by a small implant base, was not typical of urothelial cancer, and led us to opt for a conservative treatment. Therefore, we performed multiple diagnostic biopsies with laser-ablation of the lesion.

Considering that malakoplakia is rare, no standardized treatment or follow-up suggestions are available. General principles of treatment include systemic antibiotics, surgical excision, and limiting immunosuppression. Surgical excision of malakoplakic lesions should be considered based on the site affected, possible complications, or medical treatment failure.<sup>1,4</sup> Malakoplakia of the upper urinary tract tends to be aggressive. In fact, even though it is considered a benign condition, malakoplakia can clinically mimic malignancy and cause ureteric obstruction, resulting in acute renal failure and kidney damage.<sup>2</sup> Therefore, a timely and effective intervention is necessary to avoid renal failure. In cases of emergency, renal drainage via ureteric stents or percutaneous nephrostomy may be required.<sup>2</sup> Accordingly, our patient presented with a distal ureteral obstruction resulting in a grade 2 hydronephrosis of the right kidney and an increase in serum creatinine.

After the histological finding of malakoplakia, a subsequent II-look uretero-rensoscopy after 2 months showed a millimetric pathological

residual tissue that was laser-ablated. We implemented a strict follow-up, with upper urinary tract imaging every 6 months, to detect early possible recurrences and avoid kidney complications. No recurrences were discovered at 12-month follow-up; the patient had no hydronephrosis and the serum creatinine was normal.

#### 4. Conclusion

Our experience suggests that, even though malakoplakia is a rare condition, it should be taken into account for an appropriate differential diagnosis, especially in women with recurrent urinary tract infections by Gram-negative microorganisms. In case of upper urinary tract involvement, when possible, a conservative endoscopic surgical approach with laser-ablation should be performed, as well as an II-look ureteroscopy should be considered, to ensure complete resection of the pathological tissue and avoid recurrences. A conservative approach allows renal function preservation, but its effectiveness should be assessed over the long term. Therefore, a long and strict follow-up should be considered.

#### Declaration of competing interest

The authors declare no conflict of interest.

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