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Case Report

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# Multifocal Malacoplakia of Urinary Bladder Involving Left VUJ and Concomitant Malacoplakia of Right Distal Ureter Presenting with Obstructive Uropathy: A Rare Case Report

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## Abstract

*Malacoplakia is a very rare chronic granulomatous disease. It can affect almost all organs of the urogenital system, among which the urinary bladder is most common. During cystoscopy, it may reveal sessile mass which may cause misdiagnosis as malignancy. Therefore, only histopathological examination can differentiate it from malignancy. We reported a case of a 31 years old male presented with generalised weakness, low grade fever, dysuria, frequency and urgency but no pyuria or haematuria. Diagnostic urethrocystoscopy was performed which revealed multiple sessile mass obscuring left ureteric orifice, but right ureteric orifice was identified properly and right sided URS was done & soft plaque like lesion was found at distal ureter from which biopsy was taken. Then TUR of bladder mass was done resulting visualization of left ureteric orifice, followed by left sided URS in which whole of the left ureter was found dilated upto renal pelvis with no mass lesion. Microscopic examination of tissue taken by both transurethral resection and URS biopsy revealed features of malacoplakia. Postoperatively, the patient was treated with broad spectrum antibiotics and other supportive measures to prevent further deterioration of renal function. Ultimately, the patient recovers well with no more progression to obstructive nephropathy. Urologists should emphasize multifocal malacoplakia involving more than one genitourinary organ presenting with obstructive nephropathy. Early diagnosis and treatment with long term oral antimicrobial agents combined with cholinergic drugs or vitamin C, as well as full surgical excision (i.e. transurethral resection) should be done to obtain better therapeutic effect against malacoplakia.*

## Introduction

Malacoplakia is derived from the Greek word malakos, "soft" and plakos, "plaque" - meaning "soft plaque". It is an unusual chronic granulomatous inflammatory disease which was originally described by Michaelis and Gutmann. Most common sites of malacoplakia are the genitourinary system, gastrointestinal tract, retroperitoneum, skin etc. In urinary system, the urinary bladder is most frequently affected, followed by the kidney and rarely the ureters.[1]The pathogenesis of malacoplakia is thought to be due to macrophage dysfunction resulting in impairment to kill the offending bacteria. The disease most commonly occurs in chronic debilitating and immunocompromised patients.[2] Recurrent UTI with E. coli is one of the most common risk factors to develop malacoplakia. It can present as nodules, plaques, sessile mass or ulcers, but may be mistaken for a malignant tumor of the similar

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organ. The radiological findings of the disease are not different from those of malignant tumors. So, there is high risk of misdiagnosis of malacoplakia. However, histopathological examination and immunohistochemistry can confirm the diagnosis of the disease. In the recent years, malacoplakia has been reported worldwide and satisfactory results are obtained after proper treatment. We have reported a 31 years old male with histopathologically confirmed diagnosis of malacoplakia of urinary bladder invading left ureteric orifice and concomitant malacoplakia of right distal ureter resulting bilateral hydroureter-onephrosis and obstructive nephropathy which is very rare and first time seen in Bangladesh.

**Case report:**

A 31 years old male was presented with generalised weakness, low grade fever, dysuria, frequency and urgency. He suffered from recurrent UTI but was non-compliant with adequate treatment. There is no history of taking steroids or immunosuppressive drugs. He was non-smoker, non-alcoholic and non-diabetic. None of his family members suffer from such kind of illness. On the physical exam, he was hypertensive. Body built and nutritional status were within normal limits.

The results of laboratory analysis were as follows: haemoglobin - 10.3 gm/dl, erythrocyte sedimentation rate - 81.00 mm in 1st hour; serum creatinine - 3.53 mg/dl; eGFR – 23 ml/min; serum electrolytes: sodium - 132 mmol/L, potassium - 5.1 mmol/L, chloride - 101 mmol/L. Urine analysis showed pyuria, microscopic haematuria, proteinuria and urine culture revealed growth of E. coli. Non-contrast CT scan of KUB region revealed thick, trabeculated & sacculated wall of urinary bladder; dense area in right terminal ureter and bilateral gross hydroureteronephrosis (**figure 1**).

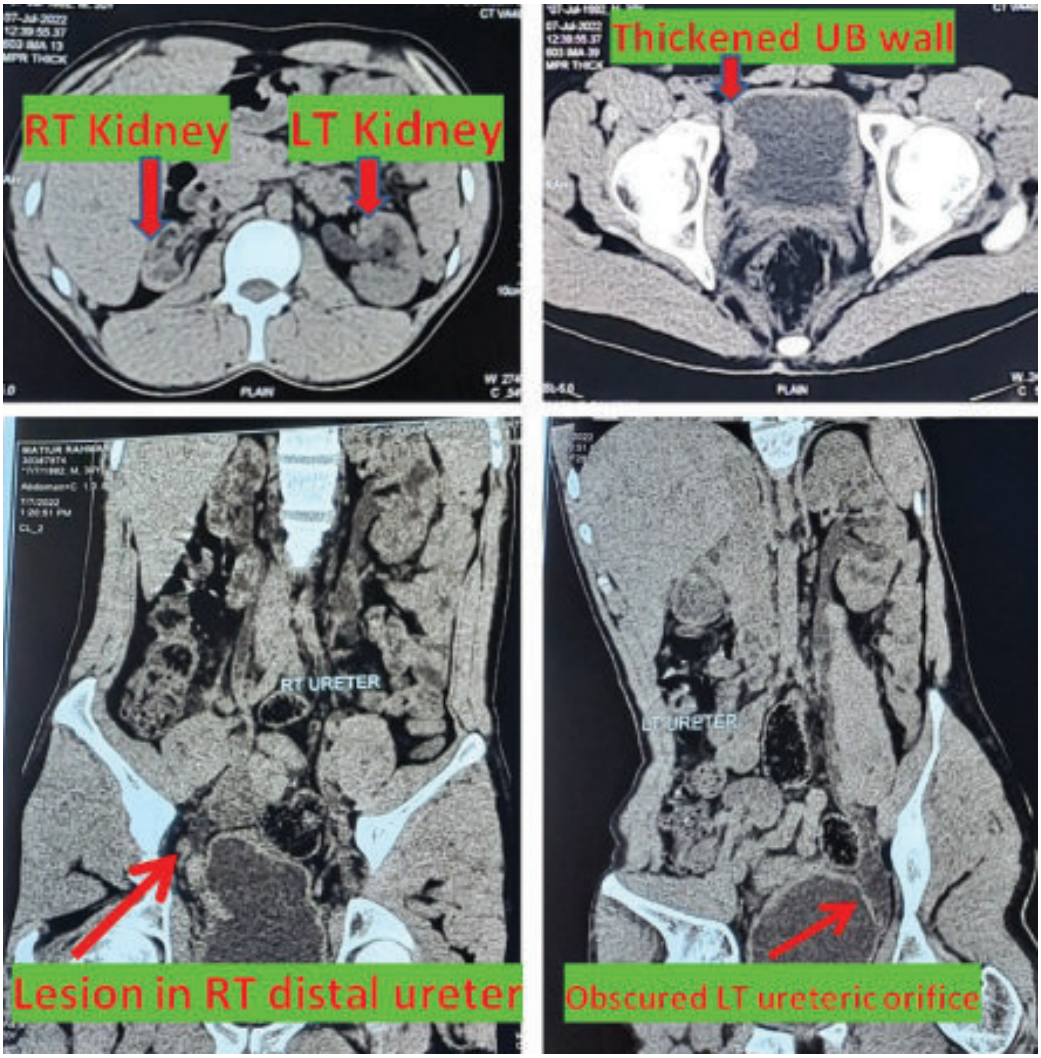


**Figure 1**: *NCCT scan of KUB region showing a thickened, sacculated and trabeculated urinary bladder wall, a dense lesion in the right terminal ureter and bilateral gross hydroureteronephrosis.*

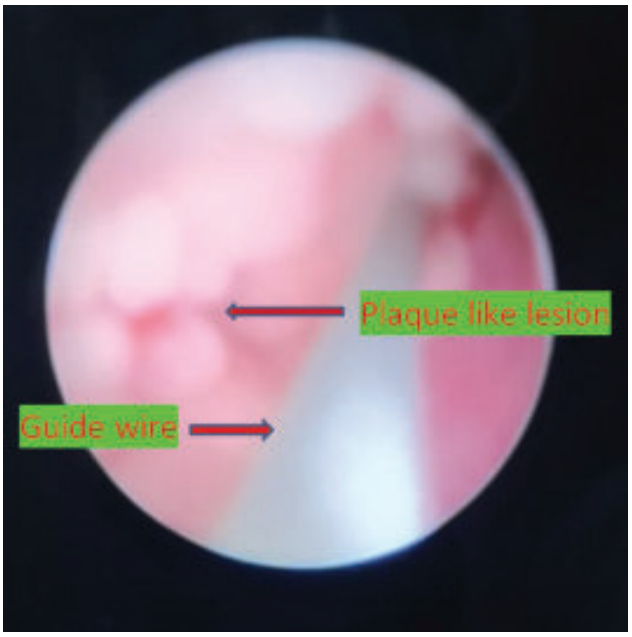


**Figure 2**: *Malacoplakia of right distal ureter seen as plaque like lesions during URS.*

After taking informed written consent, the patient underwent urethrocystoscopy which revealed multiple foci of sessile mass within urinary bladder involving left lateral wall, posterior wall, base of bladder and very close to left VUJ that obscured left ureteric orifice. Right ureteric orifice was identified properly and URS was done that revealed plaque like lesions at the right distal ureter (**figure 2**) from which the biopsy was taken. Then TUR of bladder mass was done resulting in visualization of left ureteric orifice. Finally, left sided URS was done and whole left ureter was found dilated upto renal pelvis with no mass lesion.

Histologically, on hematoxylin and eosin (HE) staining, tissue taken by both transurethral resection and URS biopsy showed infiltration of eosinophilic chronic inflammatory cells including histiocytes & lymphocytes with the presence of abundant basophilic Michaelis-Gutmann corpuscles which were positive in PAS stain and Perls Prussian blue stain (**Figure 3)**.

Postoperatively, the patient was closely monitored and perurethral catheter was removed on 7th post-operative day. During discharge, the patient was advised to continue oral antibiotic and regular follow up with ultrasonography of KUB region with MCC with PVR, urine R/M/E & C/S, serum creatinine, serum electrolytes and urethrocystoscopy every 1-3 months.

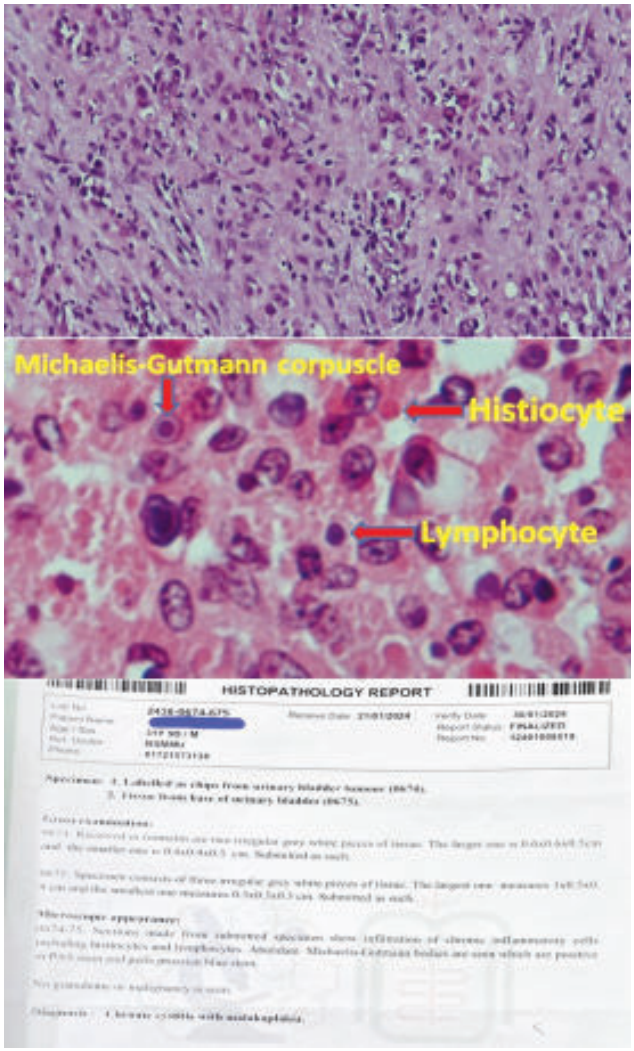


**Figure 3:** *Histopathological findings of malacoplakia of urinary bladder*

## Discussion

Malacoplakia of the genitourinary tract is an extremely rare chronic inflammatory disease. It can affect both men and women, with a male : female ratio of about 1:4. Most common age groups are over 40 years, but children or adolescents may be affected. The incidence of malacoplakia is highest in the genitourinary system in which urinary bladder malacoplakia accounts for about 40% and ureter is involved for about 11% cases.[3] Patients may suffer from immunodeficiency syndrome, autoimmune disease, HIV infection, alcoholic liver disease, tuberculosis, diabetes mellitus, sarcomatosis or other systemic disorders. Malacoplakia should be suspected in a post-renal transplant recipient who suffers from persistent UTI in spite of being treated with adequate antibiotic therapy. The disease may also

be associated with inflammatory bowel disease which supports the theory that the malakoplakia is the ultimate fate of chronic inflammation and dysregulation of the immune response.[4]

The pathogenesis of malacoplakia is unknown, but several theories are postulated.[5] There is a well-established relationship between coliform bacterial infections, mainly E. coli, and immunosuppressive diseases to develop malacoplakia. Most clinical researchers support theories that a defect in the phagolysosomal activity of the monocytes and macrophages is responsible for the unusual immunological response causing malacoplakia. It is hypothesised that bacteria or bacterial fragments form the nidus for the calcium phosphate crystals that laminate the Michaelis-Gutmann corpuscles.

The clinical presentations of malacoplakia differ from organ to organ of the body. It may lead to chronic inflammation for a long period of time. Malacoplakia of the urinary bladder may be presented as frequency, urgency, dysuria and occasional gross or microscopic haematuria. Cystoscopically, malacoplakia may be found as multifocal sessile mass lesions mimicking bladder tumours. Moreover, if ureter is involved, it may cause hydroureteronephrosis leading to obstructive nephropathy, as we presented in this particular case.

The imaging characteristics of malacoplakia are nonspecific and most of the affected organs develop tumor-like sessile mass lesions.

The diagnosis of malacoplakia mainly depends on histopathological findings with lesions being mostly found under the mucosa.[6] The lesion is characterized by infiltration of chronic inflammatory cells that includes lymphocytes, plasma cells, macrophages as well as large histiocytes, known as von Hansemann cells and small basophilic, extra or intracytoplasmic calculospherules called Michaelis-Gutmann corpuscles, which are pathognomonic for diagnosis of malacoplakia, but it may not be identified in early malacoplakia. Michaelis-Gutmann corpuscles were positive in special staining with Periodic acid–Schiff and Perls Prussian blue stain and our patient was diagnosed as malacoplakia according to the histopathological findings.

Bladder malacoplakia typically shows three stages of histopathological changes: (1) the early inflammatory stage shows infiltration of chronic inflammatory cells in the bladder mucosa, (2) the second stage is that of typical granulomatous changes showing Michaelis-Gutmann corpuscles, macrophages and occasionally giant cells & lymphocytes, (3) the last healing stage reveals fibroblasts and collagen gathering around macrophages, with simultaneous persistence of Michaelis-Gutmann corpuscles.[5]

Differential diagnosis of malacoplakia may include other granulomas, tuberculer nodules, primary & secondary malignant tumours, but no Michaelis-Gutmann corpuscles are demonstrated during histopathological studies of these diseases.[7]

There are no evidence based studies to manage the malacoplakia. The current treatment guidelines mainly aim to maintain the sterility of urine. According to etiopathogenesis of malacoplakia, antimicrobial treatment with quinolones, sulfamethoxazole and rifampicin etc. may control the disease. It has been postulated that cholinergic drugs (chlorocholine, aminoacylcholine) and vitamin C improve the defective phagocytosis of macrophages and enhance immunodeficiency state of the patients by reducing the oxidative stress and increasing the cGMP/cAMP ratio. The synergistic action of these two drugs and antimicrobial agents can significantly improve cure rates of malacoplakia.[8] If medical treatment fails, surgical intervention may be needed, together with postoperative antibiotic therapy. The duration of drug therapy is dependent on the follow up results of the disease and can be shortened when combined with surgical treatment. The long term prognosis of malacoplakia seems to be related to the extension of the disease in single or multiple organs of the body.

In this rare case, we performed transurethral resection and antibiotic treatment after diagnosis of malacoplakia. Immediately after surgical resection, renal function improves dramatically and the patient gets rid of the hallmark of obstructive nephropathy. Therefore, we believe that transurethral resection was rational to enhance the effectiveness of treatment of multifocal malacoplakia of the urinary bladder involving ureteric orifice.

**Conclusion**

Proper treatment & control of UTI is mandatory to prevent malacoplakia of the urinary system so that unnecessary surgical interventions can be avoided. Malacoplakia near the ureteric orifice can lead to obstructive nephropathy, making surveillance

cystoscopy essential in patients with chronic UTI, especially when standard treatments fail, and there is concern for underlying pathology.

**Declaration of interest**

The authors declare no conflict of interest.

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