

An Unusual Presentataion of Malakoplakia - SynchronousAffliction of Ureter and Bladder

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ABSTRACT

Malakoplakia is an unusual inflammatory disease known to affect the genitourinary and gastrointestinal tracts, skin, lungs, bones, and mesenteric lymph nodes. Malakoplakia of the bladder and ureter is quite rare. A 26 year old male patient presented with complaints of right flank pain and history of urgency and frequency since 6 months and was diagnosed to have recurrent urinary tract infections. He was evaluated and found to have persistent right hydroureteronephrosis, multiple ureteric strictures. Bladder biopsy showed evidence of malakoplakia. Patient was initially treated with percutaneous nephrostomy and later by right double 'J' stenting. Post operative follow up showed no evidence of hydronephrosis and he was relieved of his complaints. This rare case of urinary bladder and ureter malakoplakia in a young male is presented to emphasize up on the fact that malakoplakia can still occur in young male with recurrent urinary tract infection especially in developing countries like India.

Keywords: Malakoplakia, right double 'J' stenting, ureter, bladder

INTRODUCTION

Michaelis and Gutmann first described malakoplakia in 1902^[1], Malakoplakia is a Greek word malakos means soft, and plakos means plaque.^[2] The age at diagnosis ranges from 6 to 85 years, with an average age of 50 years at presentation. There is a female predominance, with a female to male ratio of 4:1. ^[3] Malakoplakia is a chronic inflammatory disease that affects the genitourinary tract with a special affinity for bladder, typically occurs in chronic debilitated, immunocompromised, and those who have other chronic diseases, ureter is rarely involved in disease process, kidney is commonly involved but typical hydronephrosis is absent.

The exact pathogenesis is unknown, but it is generally assumed that a combination of chronic bacterial infections in a patient with chronic debility or immunosuppression causes this disease. Nearly 90% of the patients have coliform urine infections and 40% have autoimmune

disease or some type of immunodeficiency.^[4] Witherington et al have hypothesized that diminished monocytic bactericidal activity against E. coli is responsible for the unusual immunologic response that causes malakoplakia.^[4] If ureter or renal pelvis is involved the patient will manifest symptoms due to upper urinary tract obstruction. In cases of renal parenchymal infection, the patient will have fever, flank pain, and a flank mass in association with urinary tract infection. Malakoplakia of the testis may manifest as epididymo-orchitis. Prostatic malakoplakia may manifest as a hard induration on DRE mimicking carcinoma prostate.

Definite diagnosis is made by biopsy. Microscopically, there are aggregates of large mononuclear phagocytes - the von Hansemann cells admixed with intracellular and extracellular Michaelis-Gutmann bodies. Michaelis-Gutmann bodies are pathognomonic of malakoplakia and are discrete, sharply demarcated intracellular or extracellular 'calculospherules' usually with a concentric

owl-eye appearance.^[5] However, they may not be seen in the early stages of the disease and are not absolutely necessary for the diagnosis. The treatment of malakoplakia depends on the extent of the disease and the underlying conditions of the patient. The initial treatment of malakoplakia consists of prompt treatment of urinary infection and surgery for the affected site.

A cholinergic agonist, bethanechol chloride, is also used to supplement antibiotic regimens to correct the lysosomal defect after the initial treatment with antibiotics, if there is a persistence of plaques in the bladder, they can be endoscopically resected and cystoscopically followed up.

Focal involvement of only lower ureter can be treated with local excision and ureteroneocystostomy. Unilateral renal or upper tract involvement needs nephrectomy and has good results. The prognosis of bilateral renal involvement is poor with 100% mortality within 6 months irrespective of the treatment chosen.

We hereby present an unusual case of a young male diagnosed as malakoplakia of urinary bladder and ureter with persistent hydroureronephrosis, which resolved on Double 'J' (DJ) stenting. Patient on follow up was relieved of his complaints. We also highlight the pathological aspects as histopathology is important in establishing diagnosis.

CASE REPORT

A 26 year old male patient presented with complaints of right flank pain since 6 months which was intermittent colicky, dull aching. Patient had history of urgency and frequency, dysuria since 6 months, frequency D/N 10-15/4-5, there was no history of fever, no history of poor stream, intermittency, straining at voiding, and no history of hematuria, pyuria, and no co morbidities. On examination vitals were normal; per abdomen was soft, non tender, bowel sounds were heard normally. For these complaints he went to local hospital where he was evaluated.

Ultrasound abdomen showed right moderate hydroureronephrosis, ureteric calculus. CT Scan abdomen and pelvis showed right moderate hydroureronephrosis with no evidence of pyonephrosis.

Urine culture showed E.coli; urine AFB was negative, Hb-13.5gm% TLC-6900 DLC N-65.8 L-31.8 M-2%. On cystoscopy bullous oedema of bladder mucosa was present, bladder biopsy was done and right DJ stent placed. They suspected genito-urinary tuberculosis(GU TB) and ATT was started empirically but Bladder biopsy was negative for GU TB, Urine PCR- negative, patient

was discharged with right DJ stent insitu. Bladder biopsy was reported as malakoplakia, right DJ Stent was removed after 20 days.

Patient again had right flank pain 3 weeks later for which he was evaluated at a private hospital, found to have persistent right hydroureronephrosis. Intra operative findings- right DJ stent attempted, but failed, hence right PCN (Percutaneous nephrostomy) placed.

Table 1 : Showing the Investigations done

Blood	
Haemoglobin; PCV	11.7gm/dl; 37.5
Rbc count	4.62mill/cu mm
Wbc	4900cells/cu mm
DLC	N- 58%, E-1%, L-33% M-8%
ESR	20mm/1st hr
RBS	97mg/dl
Platelet count	1.61 lakhs/cu mm
Peripheral smear	Normocytic, normochromic
RFT	170.98
SE	141/3.7
HIV,HBsAg,HCV	Negative
Blood grouping and typing	B +ve
BT/CT/PT/APTT	3/6.3/14.2/32.2
Urine	
Routine	Alb-trace, sugar-nil, Pus cells-10-12/hpf Epithelial cells-4-6/hpf
Culture	E.Coli (sensitive to Ciprofloxacin Trimethoprim-sulfamethoxazole)

Patient was on ATT for 6 months , then patient presented to us with residual right HDUN and right PCN insitu. Investigations done at our institute.

DISCUSSION

Malakoplakia involving kidneys, ureters, and prostate is less common. In a review of 153 cases of malakoplakia in 1981, Stanton and Maxted found that only 11% had ureteral involvement.^[6] So far only nine cases have been reported with majority of them from Japan having involvement of ureter only without kidney being involved in the disease^[7] after that there has been only one case report of isolated ureteral malakoplakia from India. It is possible that many cases are being missed either because the clinicians are not looking out for this entity or the histopathologists are not trained to diagnose malakoplakia. In fact only about 10% of the pathologists could diagnose malakoplakia as seen in the review by Stanton and Maxted .

An Unusual Presentataion of Malakoplakia

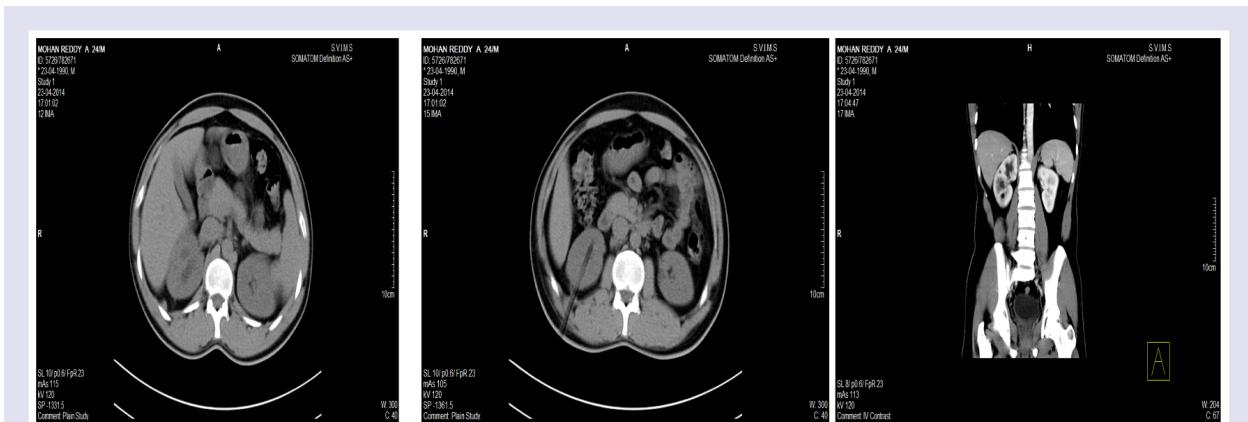


Figure 1: CECT abdomen and pelvis showing evidence of mild PCS (pelvicaliceal system) prominence with right PCN insitu, normal opacification of proximal 4.5cms of right ureter. Then patient was subjected to right nephrostogram

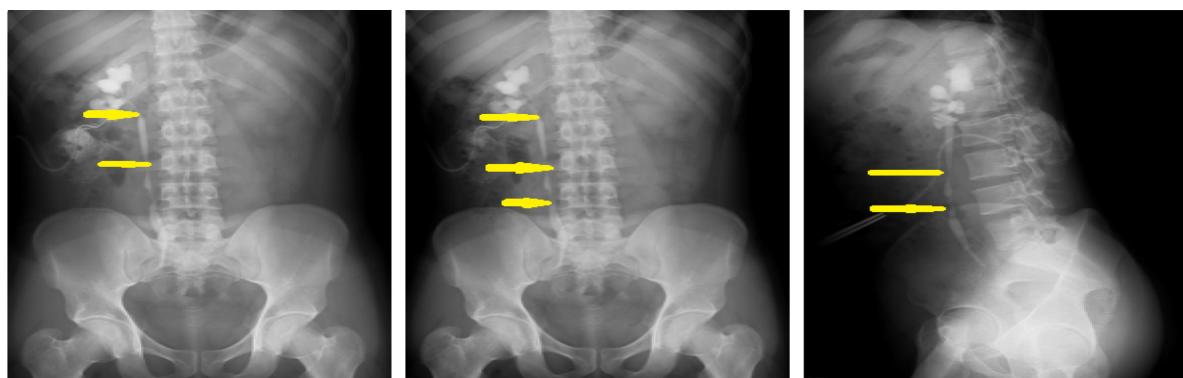


Figure 2: Right nephrostogram showing mild HDUN (hydro ureteronephrosis) with multiple short segment strictures along upper, mid and distal ureter.

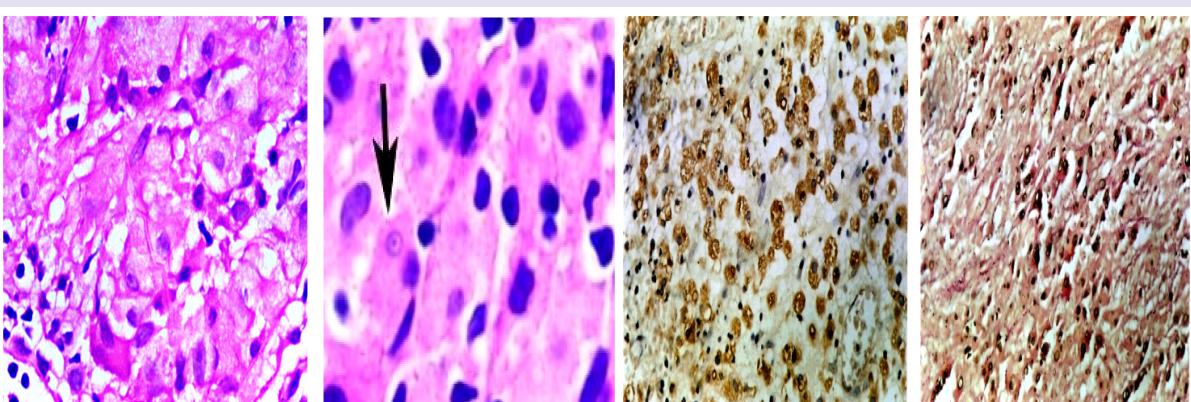


Figure 3: Slides of bladder biopsy done outside were reviewed slides (upper left) showing large histiocytes, known as von Hansemann cells (upper right) showing Pathognomonic; Small basophilic, extracytoplasmic, or intracytoplasmic calculospherules called Michaelis-Gutmann bodies (lower left) showing Macrophages with positivity for CD68 (lower right) showing Small basophilic, extracytoplasmic, or intracytoplasmic calculospherules called Michaelis Gutmann bodies (Von kossa stain). Patient was posted for surgery, initial plan was to attempt right DJ stenting If fails to go for ileal ureteric replacement.

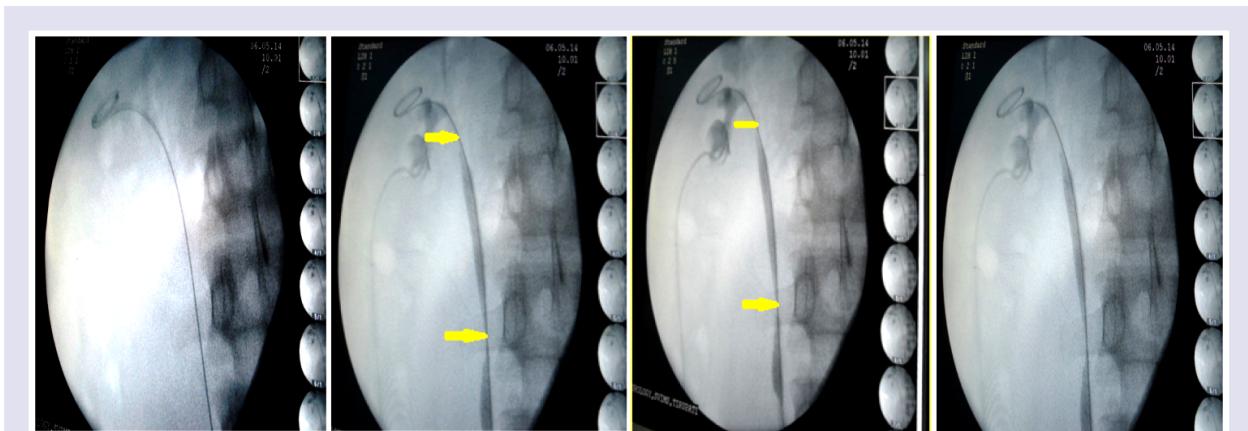


Figure 4 : Intra operative findings were turmo guide wire passed [upper left], Retro grade pyelogram done [upper right], Multiple ureteric strictures noted along the course of ureter [lower left], Double 'j' stent in place [lower right]
Patient was placed on Culture based antibiotics like ciprofloxacin, trimethoprim-sulphamethoxazole and on Ascorbic acid and Bethanechol

In the previous case of ureteral malakoplakia which has been reported Jayesh et al IJU^[8] is a girl of 15 years with distal ureteric stricture with left gross hydronephrosis for which initially dj stenting attempted but guide wire could not be negotiated hence ureteric reimplantation was done excised specimen showed evidence of malakoplakia.

There are various case reports involving urethra, kidneys and bladder but involvement of ureter is quite rare. Even more rare is the presentation of the present case he is young male majority of the reports were of women and usually elderly; present case had no comorbidities where as the other ones had either history of diabetes or usage of immuno suppressive drugs, site of involvement also is distant with both bladder and ureter involved, and also there is presence of hydronephrosis in the present case but without any kidney involvement.

CONCLUSION

Malakoplakia even though rare should be considered in the differential diagnosis of any patient with fever of an unknown origin, flank pain, history of recurrent urinary tract infections with a renal mass especially so in an immuno compromised patient.

CONFLICT OF INTEREST

The authors declared no conflict of interest.

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