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Case Report – Urologic Oncology

Reviving Bladder: Conquering Malakoplakia through Partial Cystectomy

Mesaneyi Yeniden Canlandırmak: Parsiyel Sistektomi ile Malakoplakinin Üstesinden Gelmek

Short Title: Reviving Bladder (Mesaneyi Yeniden Canlandırmak)

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Abstract

Malakoplakia is a rare chronic inflammatory disease, often affecting the genitourinary system and mimicking malignancy due to tumor-like lesions. This case involves a 40-year-old woman with painful urination after a prior urinary tract infection. Imaging and cystoscopy revealed a necrotic bladder mass, with surrounding omental inflammation and adherence to the ileum. Histopathology identified hyperplastic stratified squamous epithelium with ulceration and reactive atypia, but no evidence of granulomas or malignancy. Laparoscopic partial cystectomy revealed dense inflammatory adhesions, and the bladder mass was successfully resected.
Keywords: cystectomy, inflammation, malacoplakia, urinary bladder, urinary tract infection

Özet

Malakoplaki, sıklıkla genitoüriner sistemi etkileyen ve tümör benzeri lezyonlar nedeniyle maligniteyi taklit eden nadir bir kronik inflamatuar hastalıktır. Bu vaka, daha önce geçirilmiş bir idrar yolu enfeksiyonundan sonra ağrılı idrara çıkma şikayeti olan 40 yaşında bir kadını içermektedir. Görüntüleme ve sistoskopi, çevresinde omental inflamasyon ve ileuma yapışıklık bulunan nekrotik bir mesane kitlesi ortaya koymuştur. Histopatoloji, ülserasyon ve reaktif atipi ile hiperplastik tabakalı skuamöz epitel tespit etmiştir, ancak granülom veya maligniteye dair bir kanıt yoktur. Laparoskopik parsiyel sistektomi yoğun inflamatuar yapışıklıklar ortaya koymuş ve mesane kitlesi başarıyla çıkarılmıştır.
Anahtar kelimeler: sistektomi, inflamasyon, malakoplaki, mesane, idrar yolu enfeksiyonu

Introduction

Malakoplakia, derived from the Greek term for "soft plaque", [1] is a rare chronic inflammatory disease first identified by Professor von Hansemann in 1901 and reported by Michaelis and Guttman in 1902. Although benign, it frequently resembles malignant carcinomas due to its tumor-like mass formations, complicating its diagnosis. Depending on the location, the disease typically manifests as raised, grey lesions of varying sizes or soft, yellow mucosal plaques during physical examination [2].

Though malakoplakia can affect multiple organs, it primarily targets the urinary system, especially the bladder, with less frequent involvement of the kidneys and ureters [1].

Malakoplakia in the urinary system can lead to acute kidney injury, frequent urinary tract infections (UTIs), and renal failure, but is rarely fatal [3].

The symptoms vary depending on the affected organ: in cases involving the urinary tract and/or bladder, patients may experience frequent urination, urinary urgency accompanied by vague discomfort, hematuria, and bladder irritability; in instances of renal and ureteral involvement, symptoms can include lower back pain and fever [1,4].

The exact cause of bladder malakoplakia is poorly understood, but its pathogenesis is based on three primary hypotheses. The first hypothesis suggests bacterial infections, particularly those caused by E. coli, often occuring after a prolonged and recurrent history of chronic UTIs. The bladder's local environment fosters bacterial proliferation and triggers an inflammatory reaction in the bladder lining [5]. The second hypothesis points to immunocompromised states or long-term chronic conditions such as HIV, tuberculosis, sarcoma, diabetes, lymphoma, and ulcerative colitis [6]. Third, it is believed to stem from an acquired defect in the bactericidal function of macrophages. Normal microtubular function and phagolysosomal activity require beta-glucuronidase and cyclic guanosine monophosphate (cGMP). Reduced levels of these enzymes result in impaired clearance of pathogenic organisms due to the persistence of phagolysosomes. The characteristic Michaelis-Gutman bodies, which are calcified intracytoplasmic inclusions, represent the phagolysosomes that have failed to undergo exocytosis [3,6,7].

Malakoplakia in the genitourinary system typically shows a higher prevalence in females, with a female-to-male ratio of 4:1. The age of diagnosis can range from six weeks to 85 years, with the average age at which individuals start showing symptoms being 50 years [2,3].

Case

Informed consent was obtained from the patient. A 40-year-old female presented to the urology outpatient department with painful and burning micturition persisting for three months. Informed consent was obtained from the patient for documenting her medical data and including images in this report. She had previously been hospitalized for a UTI lasting one week and was diagnosed with a bladder mass for which cystoscopy, followed by biopsy was performed. During the cystoscopic examination, an intramural mass was seen in the fundus of

the urinary bladder. Histopathology of the biopsy specimen revealed hyperplastic stratified squamous epithelium with ulceration and reactive atypia. There was neutrophilic exocytosis with underlying stroma showing mixed inflammatory infiltrate. No atypia, granulomas, or malignancy were observed.

An MRI of the pelvis **(Figure 1)** revealed an irregular necrotic mass, approximately 3.6 cm x 2.8 cm x 4.7 cm, arising from the bladder fundus. The mass was surrounded by omental inflammation, and a short loop of ileum was adhered to its right side. Two lymph nodes were noted along the lateral pelvic wall.

During laparoscopic partial cystectomy, dense inflammatory adhesions were found between the anterior abdominal wall, bladder, and ileal loops **(Figure 2)**. The bladder mass was noted intramurally and dissected using harmonic scalpel and monopolar electrode and the two-layers were closed with continuous interlocking suturing using Triclosan coated Polyglactin 910 Trusynth plus neo suture (Healthium Medtech, Bangalore, India). The standard technique is to use absorbable sutures to close the defect and allow free catheter drainage of the bladder for at least one week. Haemostasis was achieved and post extubation, the patient was given symptomatic care. The patient was followed up on postoperative day ten and underwent Xray cystogram using urograffin dye which confirmed no evidence of leakage **(Figure 3**).

Discussion

Only 49 reports of 72 instances of bladder malacoplakia have been documented globally from 1986 to 2021. [6] A literature review by Polisini et al. included 36 case reports, of which 66.67% of patients were treated surgically. Among those, 75% underwent transurethral resection of the bladder, and 25% underwent partial or radical cystectomy. The reported recurrence rate after surgical treatment was 9.09% [8].

Malakoplakia usually has a favorable outcome but tends to recur [4]. When it occurs in the urogenital tract, it can lead to renal failure due to ureter obstruction, although the duration of symptoms and clinical presentation can vary significantly [2].

Diagnosis relies on histopathology, revealing macroscopic pseudo-tumoral formations and presence of "Michaelis-Gutman" (MG) bodies within macrophages [1]. Nevertheless, not all instances exhibit the usual pathological alterations and standard MG bodies. This is because bladder malakoplakia has three phases of pathological changes: the initial inflammatory phase

involving infiltration of inflammatory cells in the bladder mucosa; the intermediate phase featuring typical granulomatous changes, marked by the presence of MG bodies and macrophages, with sporadic giant cells and lymphocytes; and the final healing phase showing fibroblasts and collagen cells surrounding macrophages, with occasional presence of MG bodies [2,9].

The imaging and cystoscopic features of bladder chondromalacia closely resemble those of a bladder tumor, and at the time of initial diagnosis, it is often misidentified as such. It is essential to differentiate superficial bladder mucosal lesions from conditions like bladder carcinoma, adenocystitis, and other disorders [4].

CT urograms are useful for evaluating cases with multiple lesions and for assessing the effect of parenchymal and ureteral lesions on the kidneys. Renal lesions typically appear as segmental or diffuse hypodense areas, while ureteral lesions present as lacunae, both of which complicate the differential diagnosis with renal parenchymal and urothelial tumors [1]. The definitive diagnosis of this condition is established through pathological examination.

At present, there is no uniform treatment protocol available [4]. It differs based on the location of the disease, its severity, and the clinical presentation. Treatment options include antibiotic therapy with careful monitoring of lesions, nephrectomy if the renal parenchyma is compromised, or resection of obstructive ureteral lesions. In cases of ureteral blockage, renal drainage through ureteric stents or percutaneous nephrostomy becomes essential [1].

Surgical removal of malakoplakia lesions should be contemplated based on the location affected, presence of complications, or if medical intervention does not yield results. A combination of surgical intervention and quinolone antibiotic therapy may represent the most effective treatment approach [3]. Laparoscopic partial cystectomy remains an uncommon procedure often performed unplanned by the gynaecologists during other operations or for rare bladder pathologies which can be carried out effectively when indicated [10].

Conclusion

Bladder Malakoplakia, although rare, should be suspected in patients with recurrent UTIs and mass-like lesions in the genitourinary tract. As it can resemble malignancy, histopathological confirmation is essential for diagnosis. Early detection allows for effective management, often with antibiotics, though surgery may be needed for severe cases. Surgical

intervention, such as partial cystectomy, remains an effective treatment for cases where medical management fails, or when complications like obstruction or recurrent infections arise. Regular monitoring of renal function and bladder health is crucial to prevent recurrence and safeguard quality of life.

Ethics Committee Approval: N / A.
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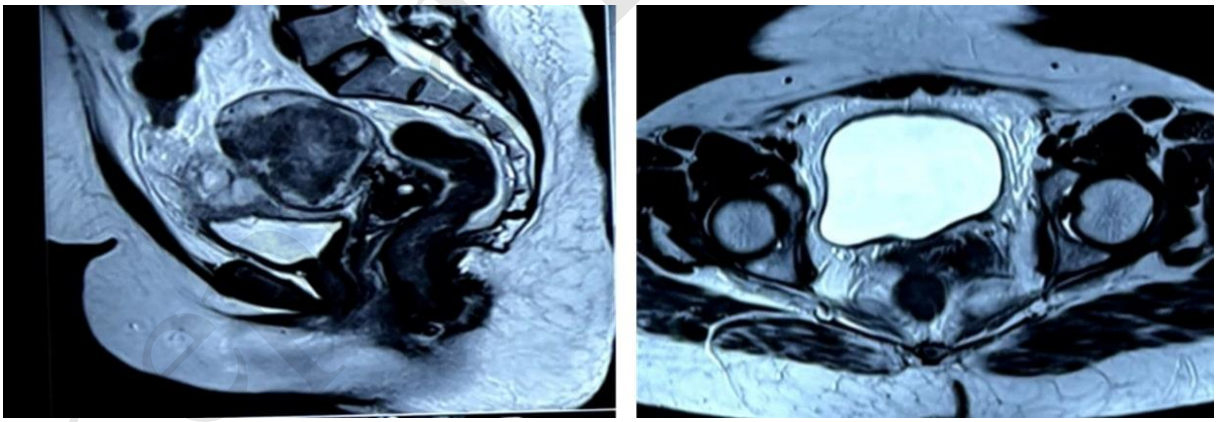

Figure 1. MRI pelvis showing irregular necrotic mass lesion arising from fundus of urinary bladder

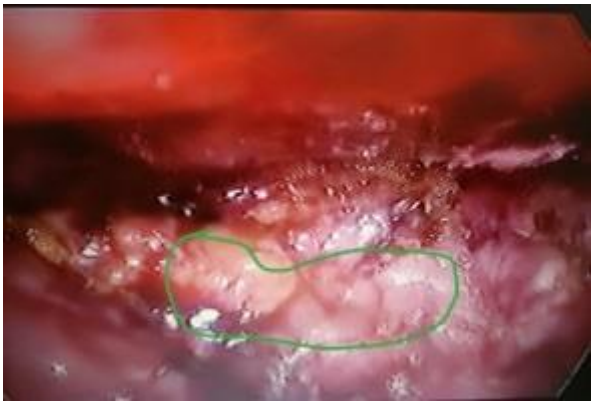

Figure 2. Adhesions between anterior abdominal wall and intramural mass (marked green)

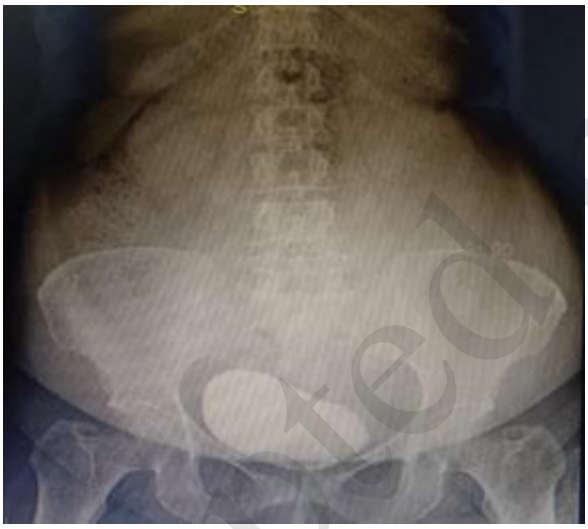

Figure 3. Xray cystogram using urograffin dye which confirmed no evidence of leakage