Isolated urachal malakoplakia mimicking malignancy

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INTRODUCTION

Malakoplakia is an unusual inflammatory disease with uncertain pathogenesis affecting any organ in the body, but predominantly genitourinary tract, with specific predilection to the bladder. We report a rare case of isolated malakoplakia of the urachus in a 29-year-old male patient who presented with lower urinary tract symptoms without any hematuria. Investigations revealed sterile pyuria with no bacterial growth in urine. Radiological investigations revealed a mass in the urachal region. The patient underwent cystoscopy with biopsy followed by pelvic lymph node dissection and partial cystectomy with excision of the urachal mass. Histopathological examination of the mass revealed malakoplakia. Postoperative course was uneventful. To the best of our knowledge, this is the first ever case report of isolated urachal malakoplakia without any concomitant malignancy or bladder involvement reported in our country and one of the very few reported worldwide.

Key Words: Malakoplakia, Michaelis–Gutmann bodies, urachal mass, Von Hansemann histiocytes

CASE REPORT

A 29-year-old male presented with suprapubic pain, dysuria, increased frequency of micturition, and intermittent episodes of fever for 8 months. No diurnal variation of fever was noted. Hematuria was absent. Loss of weight and appetite were present. Clinical examination revealed no abnormal findings. Urinalysis showed 500 leukocytes/µl; however, urine culture revealed no bacterial growth. Urine cytology was negative for malignant cells. Rest of the blood and urine examinations were within normal limits.

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limits. The patient was evaluated for genitourinary Koch’s with urine microscopy and polymerase chain reaction, which were negative for acid-fast Bacilli. Ultrasonography followed by contrast-enhanced computed tomography (CT) scan of the abdomen and pelvis was done, which revealed an irregular enhancing mass lesion with central hypodense area situated in hypogastric area measuring 68 mm × 64 mm × 50 mm in its maximum dimensions, with heterogeneity in the surrounding tissues probably due to extension or involvement of adjacent fat planes, abutting the dome of bladder with thickened bladder wall. Cystoscopy revealed no obvious intraluminal growth, with granular appearance of bladder dome mucosa with hyperemia. Biopsy of the hyperemic mucosa revealed features suggestive of cystitis cystica. The patient was explained in detail about the provisional diagnosis, surgery planned, all the complications associated with the surgery, and informed consent was duly taken. He underwent pelvic lymph node dissection, partial cystectomy with excision of mass arising from the urachus. Intraoperatively, an irregular mass arising from the urachus was noted up to the dome of bladder, with no obvious involvement of the bladder and no obvious pelvic lymphadenopathy. Partial cystectomy specimen revealed granular-appearing bladder mucosa with no obvious involvement by the mass. Final histopathology report revealed that excised urachal remnant shows features of foamy histiocytes, plasma cells, giant cells with calcium deposits, and Michaelis–Gutmann bodies, which are pathognomonic of malakoplakia. Immunohistochemistry revealed groups of numerous CD68-positive histiocytes and monocytes. Periodic acid-Schiff staining also confirmed the concomitant abundant histiocytes with Michaelis–Gutmann bodies. Bladder mucosa showed features of cystitis cystica. Lymph nodes were of reactive hyperplasia. No evidence of any malignancy was noted on histopathology. Postoperative course was uneventful and the patient was discharged with a prolonged course of ciprofloxacin and bethanechol. The patient was followed up routinely for a period of 2 years with urinalysis, ultrasonography, serum creatinine, and the follow-up was uneventful with no evidence of any recurrence.

**DISCUSSION**

Malakoplakia (also known as Von Hansemann’s disease) meaning “soft (malakos) plaque (plakos)” in Greek was first reported by Michaelis and Gutmann in 1902. It is diagnosed by its pathognomonic features of Von Hansemann histiocytes and Michaelis–Gutmann bodies. Michaelis–Gutmann bodies are intracytoplasmic or extracellular oval basophilic structures of targetoid or bull’s eye or concentric owl eye appearance consisting of mineralized (calcium phosphate crystals) undigested bacterial components trapped in lysosomes of macrophages and monocytes.

Malakoplakia is a rare inflammatory disease that affects predominantly genitourinary tract with special affinity to bladder. It also affects skin, gastrointestinal tract, lungs, and any other organs in the body. Predominantly, it is seen in males except in genitourinary tract which has a female preponderance. Usually, this disease affects patients of age more than 50 years, debilitated, and immunosuppressed patients. It is usually associated with a chronic bacterial infection. Nearly, 90% of the patients have coliform urine infections and 40% have autoimmune diseases or some type of immunodeficiency.

Malakoplakia of bladder has symptoms of bladder irritability, hematuria, dysuria, etc. Upper urinary tract

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**Figure 1:** Contrast-enhanced computed tomography scan revealing an irregular enhancing mass lesion with central hypodense area situated in hypogastric area, abutting the superior wall of bladder with thickened bladder wall.

**Figure 2:** Intraoperative findings - urachal mass without bladder wall involvement and granular mucosa of urinary bladder dome on partial cystectomy.
involvement as seen in 15% of the cases may manifest as upper urinary tract obstruction, fever, flank pain, or a mass. This patient presented with symptoms suggestive of urinary tract infection and lower urinary tract symptoms (LUTS), though bladder was found to be uninvolved, probably due to the close vicinity of the mass to the bladder. However, signs and symptoms of urachal involvement were not described in the literature, owing to its rarity in incidence.

Tian J et al. in their analysis of 33 urachal masses reported that the majority (67%) were malignant and among the benign masses (33%), only two cases were identified as malakoplakia. CT helps in distinguishing benign from malignant urachal masses. CT scan shows malakoplakia as hypodense lesions. Surgical excision is curative for benign urachal masses. Extended partial cystectomy provides a curative surgical treatment for localized urachal cancer.

Definite diagnosis is made by histopathological examination alone by demonstration of the hallmark features of malakoplakia, i.e., presence of Von Hansemann cells which are large mononuclear phagocytes along with intracellular or extracellular “calculospherules” with a concentric owl eye appearance, i.e. Michaelis–Gutmann bodies. However, they are not absolutely necessary for diagnosis as they are absent during the early stage of disease. The overlying urothelium is benign and may be hyperplastic, metaplastic, or ulcerated, but is usually intact.

Treatment of malakoplakia is primarily medical line of management with prolonged course of antibiotics such as quinolones as a mainstay and if failed, then surgery of the affected site. However, malakoplakia being mostly a diagnosis confirmed only on histopathology, in cases where it presents as a mass, mimicking malignancy, biopsy, or excision, as indicated, is advocated for both diagnostic and curative purposes. Malakoplakia of the lower urinary tract is more benign with good prognosis. The reduction in the ratio of cGMP: cAMP which affects the “redox” status of the cell, thereby affecting the lysosomal phagocytosis and causing defective microtubular assembly, was postulated to be the main defect in malakoplakia.
pathogenesis. Use of cholinergic agents such as bethanechol as a treatment had been discussed, though unproven in literature owing to its action of increasing cGMP.\(^{[12,13]}\) Similarly, ascorbic acid (Vitamin C) also may be used as a treatment owing to its property of reducing cAMP.\(^{[11‑13]}\)

We conclude that malakoplakia should also be taken into consideration as a differential diagnosis of urachal masses with solid components. Urachal malakoplakia can present as a urachal mass with LUTS without any coexisting bacterial infection, hematuria, without any concomitant malignancy or bladder involvement.

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**Conflicts of interest**
There are no conflicts of interest.

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