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Urachal Malakoplakia As A Differential Diagnosis Of Urachal Mass: A Case Report

Dr.B.V.R.S.Sai Virinchi Yadav[1], Dr. Mamidala Srinivas[2],
Dr.D.V.S.R.K.Prasad[3], Dr. G.Mallikarjuna[4], Dr. S.Anand[5]

[1]*(Post Graduate,Department Of Urology, Osmania Medical College, Telangana, India)*
[2]*(Post Graduate,Department Of Urology, Osmania Medical College, Telangana, India)*
[3]*(Professor,Department Of Urology,Osmania Medical College,Telangana, India)*
[4]*(Professor,Department Of Urology,Osmania Medical College,Telangana, India)*
[5]*(Associate Professor,Department Of Urology,Osmania Medical College,Telangana, India)*

Abstract:
Malakoplakia is a rare chronic inflammatory disease that most commonly affects the genitourinary tract. In the genitourinary tract it most commonly involves the urinary bladder. Urachal malakoplakia is rare and very few cases have been reported.We present a case of 54-year old patient who preseted to us with features of urinary tract infection and a palpable suprapubic mass. Imaging was suggestive of a urachal mass. Patient underwent en bloc resection with partial cystectomy. Histopathological examination revealed Von Hansemann cells and Michaelis Gutmann bodies without involvement of bladder mucosa and a diagnosis of urachal malakoplakia was made. Hence urachal malakoplakia needs to be considered as one of the differential diagnosis of urachal mass along with malignacy and other benign causes like cyst, abscess and xanthogranuloma

Key Word*: Urachal mass; Malakoplakia; Michaelis-Gutmann bodies,Uurachal carcinoma*

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I. Introduction

The urachus is a lower midline abdominal structure that extends from the dome of the urinary bladder toward the umbilicus, as the median umbilical ligament within the space between the peritoneum and transversalis fascia[1]. It is an embryologic fibrous remnant of the allantois, which extends between the bladder dome and the umbilicus. Defective obliteration of the urachus is rare and leads to urachal abnormalities in adulthood, most commonly masses. Majority of the urachal masses in adults are malignant**.** The benign masses include abscesses, cysts, malakoplakia and xanthogranuloma.[2]

Malakoplakia is a rare chronic inflammatory disease that was identified in 1901 by Professor von Hansemann and first reported in 1902 by Michaelis and Guttman in a urinary bladder specimen. Since then, malakoplakia has been reported in a multitude of organ systems, including the gastrointestinal tract, bones, lungs and skin[3]. It can affect any part of the urinary tract, with the bladder most usually involved, followed by the renal parenchyma, ureter and renal pelvis. [4] Involvement of urachus is very rare and very few cases have been reported. Here, we present a case of urachal malakoplakia.

II. Case Report

A 54-year-old male presented with chief complaints of dysuria, increased frequency and urgency of micturition and suprapubic discomfort. No complaints of fever or hematuria were noted. Patient did not complain of any obstructive Lower Urinary Tract Symptoms. However, loss of weight and appetite were present. Clinical examination revealed a mass in the hypogastrium approximately 6 x 3 cms in size, firm in consistency, non tender, with irregular surface and margins. Routine investigations revealed him to be a diabetic(newly diagnosed). Complete urine examination showed 500 leukocytes/µL and glucose 500mg/dl. E.coli was isolated in urine culture. Urine cytology was negative for malignant cells. Rest of the blood and urine examinations were within normal limits. Urine for Acid Fast Bacilli and Polymerase Chain Reaction test for Tubercle bacilli were negative. Ultrasonography of abdomen revealed 10.6x6.3x6.3 cm sized well defined heterogeneously hypo-echoic solid cystic lesion arising from dome of bladder , extending till umbilicus, with some areas showing internal vascularity on Colour Doppler. Rest of the organs were normal.A contrast-enhanced computed tomography (CT) scan of the abdomen and pelvis was done, which revealed a well-defined hypodense mass lesion measuring 9.8 x 6.2 x 7.2 cm, arising from dome and anterior wall of bladder, extending

till umbilicus, with punctate calcifications within it and adjacent fat stranding, closely abutting anterior abdominal wall [figure 1], no lymph node metastases were seen. Bladder wall showed irregularity at the dome.

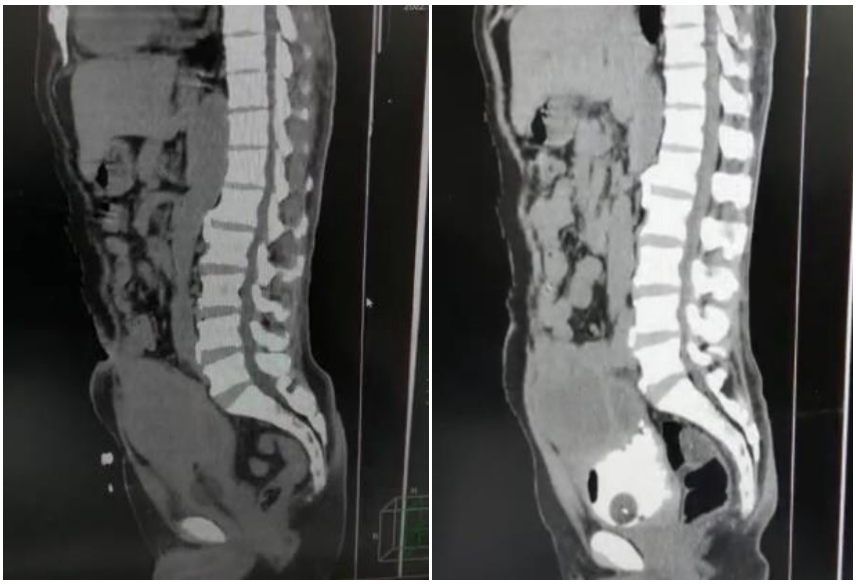

Figure 1: Sagittal section of contrast enhanced computed tomography of abdomen and pelvis showing the urachal mass at the dome of the bladder. A delayed excretory phase scan clearly shows the mass separate from the bladder with irregularity of the bladder dome.

Cystoscopy did not reveal any obvious intraluminal growth, but there was hyperemia of the mucosa at the dome of the bladder. A provisional diagnosis of lolcalised urachal carcinoma was made, and after informing the patient and taking due informed consent ,surgery was planned. An en- bloc resection of the umbilicus, urachal mass and partial cystectomy was carried out[Figure 2]. No enlarged lymph nodes were noted intra operatively.

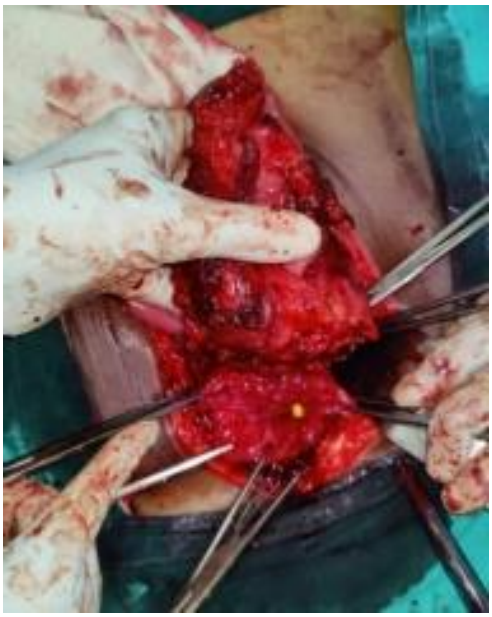

Figure 2 : En bloc resection of umbilicus, uachal mass and partial cystectomy showing urachal mass at the dome of the bladder.

Gross examination of the specimen revealed a brownish tumour extending from dome of urinary bladder to the umbilicus. On cut section cystic degeneration was seen.Partial cystectomy specimen revealed granularity of bladder mucosa with no obvious mass [Figure 3].

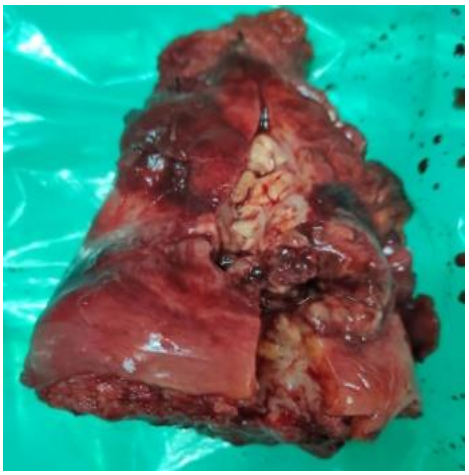

Figure 3: Gross examination of specimen revealing an irregular brownish mass with cystic degeneration

Histopathological examination revealed the excised urachal mass to show features of foamy histiocytes(von Hansemann cells), plasma cells, and Michaelis–Gutmann bodies, which are pathognomonic of malakoplakia [Figure 4].There was no involvement of urinary bladder wall

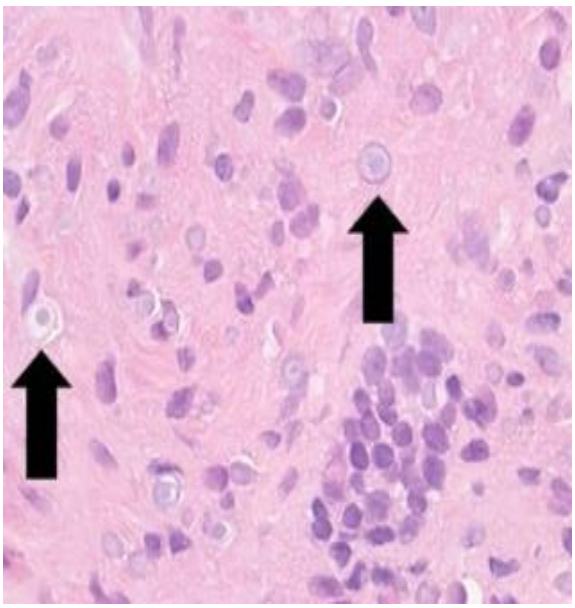

Figure 4: Histopathological Examination of urachal mass revealing foamy histiocytes, plasma cells and Michaelis Guttman bodies (black arrow marks)

Patient was discharged on day 5 and kept on a prolonged course of ciprofloxacin (as per the sensitivity report) and solifenacin for 3 months. Patient was followed up with clinical examination, ultrasound and urine examinations for a period of 1 year and the follow up period was uneventful

III. Discussion

Malakoplakia is a rare granulomatous inflammatory condition, which most commonly involves the urinary bladder and it is considered to develop as a result of a defective immune response to bacterial agents. The initial description of malakoplakia was done in a bladder specimen by Michaelis and Gutmann.[5] However, the term 'malakoplakia' was coined by Von Hansemann in 1903, which means 'soft plaque'. On gross examination a soft, yellow plaque in the bladder was noted. On microscopic examination, they identified small cytoplasmic basophilic inclusions resulting in a targetoid appearance. These targetoid cells are named after Von Hansemann and the inclusions are called Michaelis-Gutmann bodies.[6] 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. [7].

Genitourinary malakoplakia most commonly affects urinary bladder. JP Long Jr et al reported that in 9 cases of genitourinary malakoplakia in a 25-year experience, the bladder was involved in 4 patients, ureter in 2, prostate in 1, testis in 1 and a combination of prostate, bladder, rectum and pelvic adnexae in 1[8]. Involvement of urachus is however rare and very few cases have been reported.

Escherichia coli was most commonly present in urine cultures in cases of malakoplakia[8].In our case also, E.coli was isolated in urine culture.

The occurrence of urachal mass in adults is rare.Tian J *et al* reported in their analysis of 33 urachal masses that the majority (67%) were malignant and among the benign masses (33%), only two cases were identified as malakoplakia**.** [2] The urachal masses of 11 patients (33%, 11/33) were benign, including 5 cases of abscess, 3 of cyst, 2 of malakoplakia, and 1 of xanthogranuloma.

Hematuria(82%) is the most common presenting complaint of urachal cancer followed by suprapbpic pain(24%),voiding difficulties,(12%) mucosuria(9%), palpable infraumbilical mass(9%) and urinary tract infection(3%).[9] The literature on common manifestations of urachal malakoplakia is limited due to paucity of caes reported. Urachal carcinoma is diagnosed based on imaging, cytology and cystoscopy.Cystoscopy can identify a visible mass in about 80% of patients, whereas urine cytology will be positive in only 38%.[9]Urine cytology may be negative because of the extravesical location of the tumor**.**

Though our patient did not complain of hematuria, he had symptoms of suprapubic discomfort, frequency and urgency of micturition, a palpable infraumbilical mass and urinary tract infection. The finding of a urachal mass that was of heterogeneous density with punctuate calcifications on diagnostic imaging(computed tomography) and without any lymph node involvement led us to a provisional diagnosis of localized urachal carcinoma. A negative urine cytology and absence of mass in cystocopy could not rule out the diagnosis of urachal carcinoma in our case.

Treatment of malakoplakia is mostly medical with surgical intervention like endoscopic resection or surgical excision sometimes being necessary. There are no widely established guidelines for the medical treatment of malakoplakia but most approaches involve antibiotics[10] which work intracellularly such as quinolones (mainstay of treatment), trimethoprim and rifampicin to aid the defective phagolysosomal mechanism found in malakoplakia. These can be also be used long-term at low doses to prevent recurrence**.**

Management of malakoplakia entails treatment of urinary tract infection if present and surgical resection of the lesions to treat secondary complications and obtain tissue to exclude malignancy[11]**.** In our case, since a provisional diagnosis of localized urachal carcinoma was made, en bloc resection of umbilicus, urachal mass and partial cystectomy was done. Histopathological examination revealed it to be a case of urachal malakoplakia.

Recently, cholinergic drugs and vitamin C have been reported to improve the phagocytic function of macrophages and enhance immune efficacy by affecting the state of oxidative stress, as well as by increasing the cGMP/cAMP ratio. The combination of the two drugs and antibiotics has a certain effect on the treatment of malacoplakia [11,12.]

There are no guidelines for the use of antibiotics in preventing recurrence of malakoplakia—both in choice and duration. Studies suggest antibiotics targeting gram negative bacteria (*i.e.*, *E. coli*) and those that concentrate within macrophages, which may aid in the defective phagocyte function, should be favored[13, 14]. These include trimethoprim, ciprofloxacin and rifampicin [13, 14]. In our case, patient was discharged on a prolonged course of ciprofloxacin and solifenacin for 3 months.

To our knowledge, only one other case of isolated urachal malakoplakia without concomitant malignancy has been reported. [15]

IV. Conclusion

Urachal malakoplakia is a rare disorder and must be considered when a diagnosis of urachal mass is made, especially in elderly and immuno-compromised patients. Confirmatory diagnosis can be made only on histopathological examination and hence the mass needs to be resected..

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