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.

We hereby present an interesting case of a female diagnosed as malakoplakia of urinary bladder with growth. We also highlight the pathological aspects as histopathology is important in establishing diagnosis and confirmed by Special stains.

CASE REPORT

A 43-year-old female came with symptoms of pain abdomen to Yashoda Hospital, Malakpet. Routine investigations documented. All were normal including complete urine examination and ultrasound abdomen pelvis showed cystitis and small growth in urinary bladder.

Cystoscopy revealed multiple growths in the bladder, largest measuring 2X3 cms on the base, lateral wall and dome of the bladder, ureteric orifices were normal. TUR(BT) was sent.

Gross examination: There were multiple grey white soft tissue bits totally measuring 1x0.5x0.5 cms.

Histopathological Examination: Revealed urothelial lining which is focally ulcerated and focally showed squamous metaplasia, lamina propria showed sheets of many histiocytes showing abundant eosinophilic and
Figure 1: H&E showing focal ulcerated urothelium and histiocytes in lamina propria-scanner view

Figure 2: H&E showing many histiocytes in lamina propria- 10X.

Figure 3: H&E showing histiocytes with eosinophilic cytoplasm and refractile inclusion bodies-40X

Figure 4 & 5: *SPECIAL STAINS IRON & VON KOSSA: highlights michaelis gutmann bodies-40X
foamy cytoplasm. Also seen are round to oval refractile inclusion bodies reminiscent of Michaelis Gutmann bodies in cytoplasm of histiocytes. Fig (1,2,3) Inclusion bodies are concentrically laminated calcospherites highlighted by special stains iron and Von kossa for calcium. Fig (4,5)

Special stains: VON KOSSA and IRON - highlightens Michaelis Gutmann bodies.

DISCUSSION

Malacoplakia involving kidneys, ureters, and prostate is less common. In a review of 153 cases of malacoplakia 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.

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. Cystoscopically these can be confused with carcinoma. Malacoplakia 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 ‘calcospherules’ usually with a concentric owl-eye appearance. These are seen within histiocytes and interstitium. Electron microscopy shows macrophages which have phagosomes that are packed with undigested bacterial products.[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.

CONCLUSION

Malacoplakia 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 growth in urinary bladder especially in an immuno compromised patient and even without any symptoms of urinary tract infection and can be a incidental finding as in our case.

CONFLICT OF INTEREST:
The authors declared no conflict of interest

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REFERENCES