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# Mullerianosis of Urinary Bladder: A Great Impersonator of Malignant Urinary Bladder Tumours

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#### **ABSTRACT**

**Introduction:** Mullerianosis of urinary bladder is an enigmatic entity that resembles carcinoma of bladder in many aspects. However, the management of this benign condition is entirely different.

**Case report:** Herein we report a case of mullerianosis of urinary bladder with a brief review of literature to make the fellow urologist aware and more adept in dealing with this condition.

**Conclusion:** An awareness of this rare entity is necessary for adequate treatment and to the spare the patient of the stigma of a malignant disease.

**Keywords:** Mullerianosis, Urinary Bladder, Impersonator of Malignant Urinary, Bladder Tumours

#### INTRODUCTION

Mullerianosis of urinary bladder is a very rare entity comprising of admixture of Mullerian derived tissues in a urinary bladder lesion. This condition was not even known till two decades back when it was first described by Young and Clement in 1996. Less than thirty cases of this entity are described in English literature till date.<sup>1</sup>

The enigma of Mullerianosis is that it mimics a neoplastic bladder lesion clinically, cytologically and well as histologically. The rarity contributes to misdiagnosis. The correct identification of this condition is extremely important since the treatment is drastically different from malignant bladder neoplasm. While the treatment of a malignant bladder tumour invariably requires surgery; Mullerianosis often responds to hormonal manipulation and surgery is usually not required except in a few recalcitrant cases. Here we report this rare case of bladder Mullerianosis.

# **CASE REPORT**

A forty-year lady was referred to us with complaints of dysuria, frequency, urgency for period of last one year. Abdominal examination revealed mild suprapubic tenderness. On Genitourinary examination, patient had normal external genitalia with normal appearing urethral meatus. Per vaginal examination showed smooth vagina with no mass, pink, smooth cervix with no cervical discharge.

She was a known diabetic, well controlled on oral hypoglycaemic agents. There was no history of hematuria or weight loss. There was no history of tobacco use, and no known exposure to industrial dyes or other carcinogens.

Abdominal Ultrasonography showed a bladder lesion of size 4 X 3 cm on the posterior wall. Contrast enhanced Computed Tomography (CECT) showed an intraluminal bladder lesion

enhancing mass of size 5 X 4 cm arising from the posterior wall. (Figure 1) Urinary routine examination and microscopy was normal. Urinary cytology was negative for atypical or malignant cells.

A provisional diagnosis of primary bladder malignancy was made and patient was taken for cystoscopy. Cystoscopy showed a broad based solid growth on the trigone and right lateral wall of bladder (Figure 2). The mucosa overlying the lesion was intact. The ureteric orifices was seen separately away from growth. Multiple transurethral resection biopsies were taken from the growth and the specimen sent for histopathological examination.

Histo-pathological examination showed endometrial glands with focal areas of ciliated tubal epithelium (endosalpingiosis). The Mullerian glands and stroma were noted in lamnia propria and detrusor muscle. On Immunohistochemistry, the glands and stroma were immunoreactivity for both oestrogen and progesterone Receptors. CD 10 positivity was demonstrated in stromal cells (Figure 3).

The patient was started on gonadotropin releasing hormone (GnRh) analogue. Injection leuprolide 3.75 mg at three weekly intervals for four cycles was administered to the patient. The patient had good symptomatic improvement.

The patient came back one month later with recurrence of similar symptoms as previously. Despite initial improvement, the symptoms have recurred after stopping GnRh analogue therapy. The CT scan done at third month post operatively continues to show the bladder mass with no evidence of resolution.

The patient was counselled about the need for partial cystectomy in view of persisting and worsening symptoms with no response to medical therapy. However, the patient was not willing for surgical intervention. Hence the patient was given another course of four cycles of leuprolide. Patient again had symptomatic improvement. At present the patient is under close regular follow up and has been counselled about need of surgical therapy (partial cystectomy) if the

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Figure-1: CT Abdomen - Pelvis

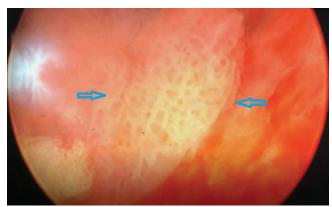


Figure-2: Cystoscopic image of lesion (Arrow denotes lesion)



**Figure-3:** Haematoxylin – Eosin stain of lesion showing endometrial (green arrow) and tubal ciliated epithelium (red arrow)

symptoms recur.

## **DISCUSSION**

Mullerianosis is a rare, benign complex tumour like lesion of the bladder comprising of Mullerian tissue in an organoid structure. It is diagnosed by presence of two of the three Mullerian tissues: endometriosis, endocervicosis and endosalpingiosis ectopic location in body. The urinary bladder is the most common site of both Mullerianosis and endometriosis. Other rare sites are ureter, mesosalpinx, inguinal nodes, spinal cord.<sup>2</sup>

The first description of this entity was given by Clement and Young in 1996.<sup>3</sup> Clement and Young had reported three cases with bladder masses on posterior bladder wall. They were treated by complete transurethral resection.

Since then, less than 30 cases have been described worldwide. It is generally seen in women between second to fifth decade of life. Patient usually presents with lower urinary tract symptoms like hematuria, dysuria, frequency, pelvic / suprapubic pain, cyclical hematuria, cyclical pelvic pain. Cyclical hematuria is seen in 20% of the cases.<sup>4</sup>

Various theories have been proposed to explain the genesis of this condition. The theory of cellular implantation has been proposed. Mullerianosis is often associated with past history of pelvic surgery or caesarean delivery. However, it cannot explain presence of Mullerianosis at distant sites like spinal cord.

The alternative theory is the metaplastic theory. It is based on metaplasia of cells along different lines (endocervical, endometrial, tubal) in response to an inflammatory insult or hormonal changes. This is supported by the fact that most lesions are on dome or posterior wall of bladder which is hormonally more receptive.<sup>5</sup>

### **CONCLUSION**

It a condition that impersonates malignant bladder neoplasms very closely – clinically, radiologically, histologically. This condition should be added to the long list of pseudoparaneoplastic bladder lesions which may be falsely mistaken as carcinoma. This is crucial for correct diagnosis and management.

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