CASE REPORT

Müllerianosis of the urinary bladder: a rare and problematic bladder tumour

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SUMMARY

Müllerianosis of the bladder is an extremely rare and highly symptomatic bladder tumour comprising of at least two types of Müllerian derived tissues. We present the case of a 59-year-old woman presenting with painful macroscopic haematuria associated with urgency, frequency and incontinence. She is a nulliparous postmenopausal woman without malignancy risk factors or previous abdominal surgery. Initially identified on ultrasound scan, she was taken for transurethral resection of bladder tumour (TURBT). Histology confirms the diagnosis. Repeat TURBT was required for non-resolution of symptoms. Despite a thorough resection, symptoms recurred within months necessitating partial cystectomy. Although a 'benign' lesion, this lesion proved a difficult condition to treat requiring partial cystectomy.

BACKGROUND

Bladder Müllerianosis is a very rare condition involving the presence of a mixture of two or more Müllerian (paramesonephric) derived tissues (endosalpinx, endometrium and endocervix) in a lesion within the bladder. It was first described by Young and Clement in 1996 and since then, there have been fewer than 30 reported cases worldwide.¹ It is characterised as a benign lesion, containing tissues of normal architecture; however, in all demonstrated cases—they are highly symptomatic, and often difficult to definitively manage without major surgery.

In this report we present the first case of Müllerianosis of the bladder in Queensland, and 29th in the world.

CASE PRESENTATION

A 59-year-old postmenopausal Philippino woman was referred by her local medical officer to our urology department with recurrent urinary tract infection, bothersome lower urinary tract symptoms including worsening dysuria, suprapubic tenderness, macroscopic haematuria, poor stream, mixed urgency and stress incontinence.

She is a nulliparous non-smoker with a body mass index 14.8 who denied industrial chemical or other carcinogen exposures.

Comorbidities included insulin-dependent type 2 diabetes mellitus, asthma, emphysema and a right total hip replacement.

INVESTIGATIONS

CT intravenous pyelogram (CT-IVP) showed a lobulated polypoidal lesion with low attenuation,

not demarcated on the non-contrast images toward the left bladder dome. The CT was unfortunately of poor quality due to streak artefact from her hip arthroplasty and needed ultrasonographic (US) and cystoscopic evaluation. US examination better exhibited a left anterior umbilicating echogenic lesion 2.8 cm×3 cm suggestive of a bladder tumour (figure 1A).

Cystoscopy under general anaesthesia revealed a partially solid and partially cystic midline bladder dome lesion that was resected using a 26Fr resectoscope and diathermy loop (figure 1B).

Unfortunately, despite thorough resection, post-operatively she suffered ongoing haematuria, increased urinary frequency and dysuria. MRI pelvis 1 month later confirmed the recurrence of the bladder mass—now measuring 31×33×22 mm (figure 1C). She returned to theatre for repeat transurethral resection of the lesion (figure 1D). Heavy vascularity and bleeding prevented a complete excision.

Histopathological analysis revealed cystic dilated spaces lined by both urothelium (figure 2B) and glandular epithelium resembling endocervical mucosa (figure 2D), as well as focal areas of ciliated cells. These ciliated cells resemble that of fallopian tissue (figure 2C). The epithelium was positive for PAX 8 (figure 2E), oestrogen, and progesterone receptors (figure 2F), confirming the diagnosis of Müllerianosis with both endocervical and tubal components (figure 2A).

DIFFERENTIAL DIAGNOSIS

Bladder wall neoplasia including but not limited to transitional cell carcinoma, adenocarcinoma, lymphoma, sarcoma and squamous cell carcinoma.

TREATMENT

She was initially treated with cystoscopic resection of the lesion. She subsequently required a secondary resection of the recurrent lesion. She will have a partial cystectomy in due course due to re-recurrence.

OUTCOME AND FOLLOW-UP

Although she improved in the 3 months following her second resection, her symptoms subsequently started to recur. She will subsequently have a partial cystectomy for definitive management of her bladder lesion, in hope of complete symptom resolution.

DISCUSSION

Müllerianosis was first described in 1996 by Young and Clement as a benign ectopic lesion with tissue

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Figure 1 (A) Preoperative ultrasound abdomen/pelvis—polypoidal lesion, (B) TURBT—cystic bladder lesion, (C) Postoperative MRI abdo/pelvis—recurrence visualised within muscularis propria, (D) Repeat TURBT—mass recurrence. TURBT, transurethral resection of bladder tumour.

Figure 2 TURBT Histopathology—(A) endocervical and tubal architecture consistent with Mullerianosis (B) urothelium and glandular tissue consistent with endocervical tissue (C) ciliated cells resembling tubal epithelium (D) glandular epithelium consistent with endocervical tissue (E) PAX 8 receptor stains positive, (F) oestrogen/progesterone receptor stain positive. TURBT, transurethral resection of bladder tumour.

of Müllerian descent.¹ Since its initial description fewer than 30 have been documented. We present the 29th case worldwide.

Theories have been posed regarding the pathogenesis of Müllerianosis, however no consensus has yet been reached.

The theory of Müllerian Metaplasia has been posed, describing the overbearing concept of cellular metamorphosis from a variety of different biochemical factors. Synonymous with the theory of coelomic metaplasia in endometriosis it describes the divergence in cellular differentiation, adopting a Müllerian tissue type. This change is thought to occur due to hormonal or inflammatory changes. This is supported by the finding that majority of described cases of Müllerianosis was found towards the bladder dome, sitting adjacent to the peritoneum—which is particularly hormone sensitive. Interestingly this condition is not observed in men. This may be a function of the circulating levels of feminising hormones.

The alternative theory is that of implantation.² A large number of patients have had a significant pelvic surgical history, giving credence to the possibility of cellular seeding. This however does not fully explain its presence in women with virgin abdomen or the presence of lesions containing multiple cell types, nor how previously described cases have demonstrated lesions within the spine, chest and lymph nodes.^{3 4}

The presentation is variable; however, the most common themes are those of painful haematuria, often cyclical in premenopausal women. Majority of cases had initial management with transurethral resection of the lesion; however, recurrence occurred in majority of cases.^{1 2 5–28} Some patients were concurrently treated with oestrogen blockade; however, its overall efficacy is not yet proven.^{8 20 22} A few patients went on to have a partial cystectomy, which in all cases has completely resolved the initial presenting symptoms.^{1 9 13 21 24}

The condition can masquerade as a malignancy; however, in essence it remains a benign condition. This being said, of the 29 cases, 1 has demonstrated endometrioid carcinoma within the Müllerian tissue—this highlights the importance of a thorough work up including tumour markers and staging imaging to exclude metastatic disease.

Learning points

- ▶ Although a benign lesion, most cases require extensive surgical resection to gain symptomatic control.
- ▶ Full resection is important to exclude adenocarcinoma within ectopic Müllerian tissue.
- ▶ Partial cystectomy may be required for complete resolution of symptoms.

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