# Supratrigonal Cystectomy and Ileocystoplasty in the Management of a Solitary Non-Malignant Bladder Lesion

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#### Abstract

Long-term use of oral cyclophosphamide is associated with cystitis and urothelial carcinomas. We report the case of a 47-year old female with Wegener's granulomatosis, who developed cyclophosphamide induced cystitis, severe lower urinary tract symptoms and upper tract deterioration. Further, she had an enlarging midline granuloma at the bladder dome. Supra-trigonal cystectomy was a reasonable option as the patient has an increased risk of malignancy, but no proven malignant change. In addition this approach allows the complete pathologic staging of the primary lesion including the potential for extended pelvic lymph node dissection, while preserving urinary and sexual function. Further, supra-trigonal cystectomy will improve the patient's quality of life by reducing her lower urinary tract symptoms, and decreasing the risk of upper tract deterioration secondary to high pressure voiding. The patient will require ongoing surveillance of her upper and lower urinary tract to ensure there is no future malignant change.

## INTRODUCTION

Wegener's granulomatosis is a necrotising granulomatous vasculitis that affects multiple organs including the respiratory and renal tracts. The bladder granulomas are typically midline, and thus have been referred to as "midline granulomatosis". Long-term, low-dose cyclophosphamide is the standard of care<sup>1</sup>. Cyclophosphamide, an alkylating agent, possesses potent immunosuppressive activity. Celldivision is impaired by the prevention of cross-linking of DNA strands thus decreasing DNA synthesis. Macroscopic haematuria is a common complication of cyclophosphamide due to cystitis or urothelial malignancy. The incidence of urothelial malignancy with low dose cyclophosphamide is 5% at 5 years, and 16% at 15 years<sup>2</sup>.

### CASE SUMMARY

A 47-year old female was referred to our tertiary institution with complications of Wegener's granulomatosis. She had been managed with oral methotrexate and low dose cyclophosphamide for 15 years, and had developed significant lower urinary tract symptoms which were worsening. Her symptoms included recurrent haematuria, urinary tract infections, urinary frequency, urgency and incontinence. Investigation with a CT pyelogram, demonstrated bilateral hydroureteronephrosis (See Images 1 & 2 below).

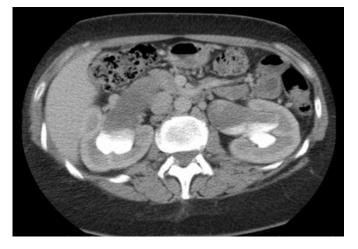
#### Figure 1

Image 1: CT pyelogram (arterial phase) with gross hydronephrosis and hydroureter (see arrow)



### Figure 2

Image 2: CT pyelogram

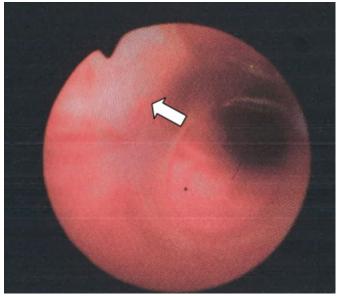


At cystoscopy bladder volume was markedly reduced and there was an ulcerative lesion at the dome of the bladder. Urine cytological evaluation was normal. Multiple bladder biopsies demonstrated a proliferative non-specific inflammatory lesion with no evidence of malignancy. Video urodynamics confirmed a small, high pressure bladder with bilateral reflux. Consequently clean intermittent selfcatheterisation was initiated.

Despite optimal technique the patient suffered recurrent urinary tract infections requiring hospitalisation. Further, the symptoms of urgency, frequency and urge incontinence became intractable. On follow-up cystoscopy, the ulcerative lesion had become thickened, indurated and clinically suspicious (Image 3).

### Figure 3

Figure 3: proliferative lesion in dome of bladder



This complex case had many factors to consider including optimum management the bladder lesion, resolving the lower urinary tract symptoms and preserving renal function. As the lesion was not malignant a radical cystectomy was not deemed necessary. Thus partial (supra-trigonal) cystectomy was performed with a detubularised small bowel neobladder to increase volume. The advantage of this technique was to improve post-operative continence and sexual function.

At operation, a lower midline laparotomy was performed. The urachus was identified and resected down to the dome of the bladder. After lateral mobilisation of the bladder and placement of stay sutures, a transverse incision was made on the anterior bladder wall. The trigone was identified and bilateral ureteric stents placed under direct vision. Gross examination of the urothelium ensured that clear margins were achievable without damage to the ureters. A supratrigonal cystectomy was performed leaving a 5 mm cuff around the trigone and the ureteric orifii. Further a cuff of peritoneum over the dome was excised. A 45-cm segment of ileum is isolated, de-tubularised and fashioned in a "W" configuration. After re-establishing bowel continuity the detubularised segment of bowel was anastomosed to the trigone. A suprapubic catheter was placed through a separate stab incision to ensure adequate drainage. Post operative histopathological examination revealed no evidence of malignancy.

#### DISCUSSION

Supra-trigonal cystectomy is not the gold standard operation for urothelial malignancy. It is an option for patients who refuse radical cystectomy with solitary bladder tumours in which it is possible to achieve a 2 cm excision margin around the base of the lesion. Partial cystectomy confers an increased risk of tumour recurrence. Further, it is contraindicated in the presence of carcinoma-in-situ (CIS), prostatic urethral involvement, or in the presence of upper tract tumours<sup>3</sup>. However, little is known about supra-trigonal cystectomy in patients with increased malignant risk but no documented evidence of malignancy.

Treatment options for this patient included radical cystectomy with ileal conduit, radical cystectomy with neobladder, and a supra-trigonal cystectomy with neobladder. The pros and cons of each procedure were discussed with the patient. Radical cystectomy has significant implications on sexual function as it leaves the vagina shortened and can predispose to vaginal stenosis. Further, radical cystectomy impacts on continence due to damage to pelvic neurovascular supply.

An ileal conduit decreases the metabolic risks of urinary diversion, but is not the diversion of choice in an otherwise healthy patient with good hand, brain and liver function. It was also unacceptable to the patient in terms of body image. However, the patient was counselled that the potential for complications associated with neobladder formation was as high as 30%.

Consequently, supra-trigonal cystectomy is reasonable given

the increased risk of malignancy, but no proven malignant change. In addition this approach allows the complete pathological staging of the primary lesion including the potential for extended pelvic lymph node dissection, while preserving urinary and sexual function. Further, supratrigonal cystectomy will improve the patient's quality of life by reducing her lower urinary tract symptoms, and decreasing the risk of upper tract deterioration secondary to high pressure voiding. The patient will require ongoing surveillance of her upper and lower urinary tract to ensure there is no future malignant change.

We expect the upper tract dilation will resolve with decreased voiding pressure. However, this may have been difficult to appreciate had an ileal conduit been fashioned as reflux may have lead to the appearance of a dilated upper tract. In addition, supra-trigonal cystectomy does not require ureteral or urethral anastomosis, and thus it decreases the likelihood of anastomotic stenosis.

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