

# Ectopic Ureterocoele with a Single System Hypoplastic Non-Functioning Kidney: Report of a Rare Case

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**Abstract:** Ureterocoele, a balloon like dilatation of distal ureter, is commonly seen in duplex system kidneys. Only 20% of cases are simple ureterocoeles seen in single system kidneys which usually have good renal function and are usually seen in children. Here we present a case of ureterocoele in an adult who presented with lower urinary tract symptoms and intraoperatively was found to be an ectopic ureterocoele with a single system kidney which was hypoplastic and non-functioning.

**Keywords:** Single system kidney, Ureterocoele, Renal hypoplasia, Non-functioning kidney, Ureterocoele excision

## 1. Introduction

Ureterocoele, a balloon like dilatation of distal ureter, is commonly seen in duplex system kidneys. Only 20% of cases are simple ureterocoeles seen in single system kidneys. These single system ureterocoeles are commonly diagnosed antenatally or during childhood and are usually of intravesical variant with a reasonably good renal function. Here we present an adult case of ectopic ureterocoele in a single system kidney which was hypoplastic and non-functioning.

## 2. Case Report

A 25 year old male came with presenting complaint of pain in lower abdomen and history of dysuria, urinary frequency and urgency for 1 month. There was no history of fever or pyuria. Per abdominal examination was normal. An ultrasound of the abdomen revealed absent right kidney with a cystic swelling in pelvis on the right side with intravesical extension. There was compensatory hypertrophy of the left kidney. His renal function tests were normal. An intravenous pyelogram was done in which right kidney was non-visualised and left kidney showed normal contrast uptake and excretion. A filling defect was noted in bladder in the delayed films suggesting possibility of an ureterocoele (Figures 1, 2).

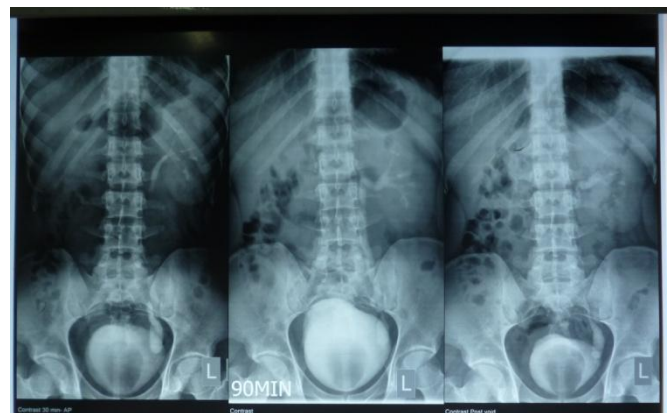


Figure 2: Intravenous pyelogram (Delayed films)

A micturating cystourethrogram also showed a filling defect in the bladder with no evidence of any vesicoureteric reflux. A contrast enhanced CT was done in which the right kidney was absent in the renal fossa, a hypodense cystic swelling was noted in the right side of pelvis with communication with bladder and intravesical extension suggestive of an ureterocoele (Figures 3,4,5).



Figure 1: Intravenous pyelogram



Figure 3: CECT showing absent right kidney



Figure 4: CECT showing ureterocele

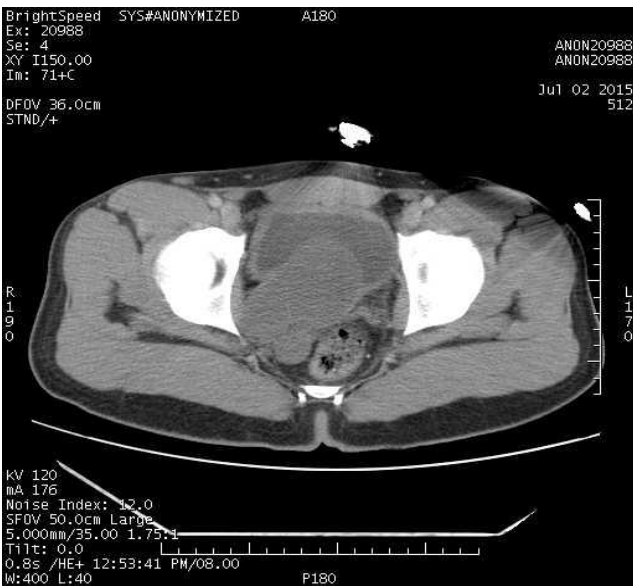


Figure 5: CECT showing intravesical extension of ureterocele

Cystoscopy showed an intravesical smooth swelling extending till bladder neck but ureteric orifice could not be made out. A DTPA scan showed no tracer uptake in renal fossa or pelvis on right side, left kidney had 100% function (Figure 6).

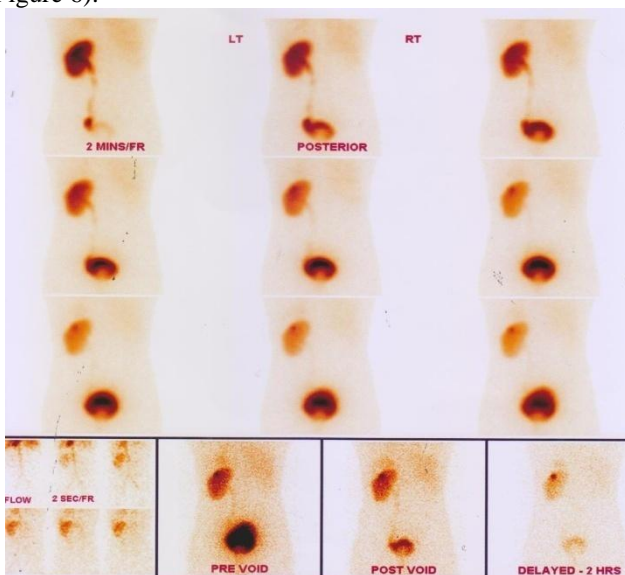


Figure 6: DTPA scan with no tracer uptake on right side

The patient was taken up for surgery in view of symptoms of abdominal pain and lower urinary tract symptoms. Intraoperatively a cystic dilatation of the right distal ureter was found communicating with the bladder and extending intravesically from posterior aspect (Figures 7, 8).

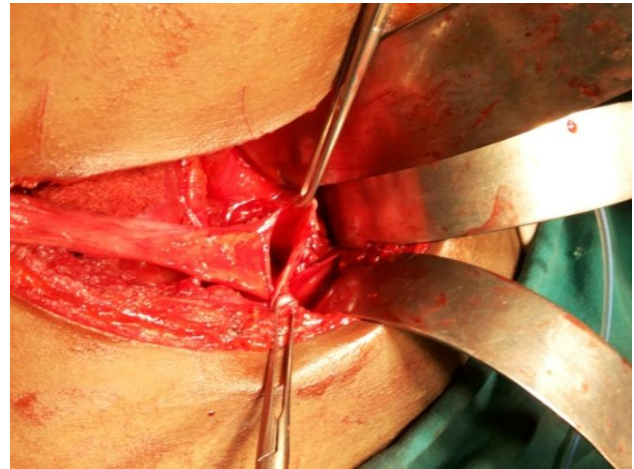


Figure 7: Intraop photo of ureter with dilated distal end

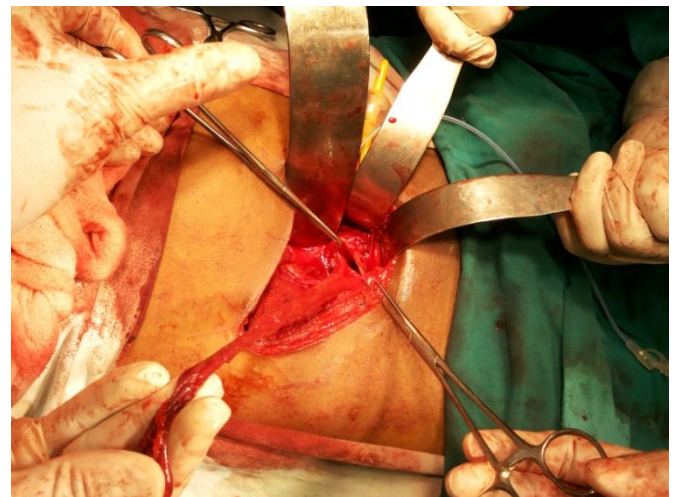


Figure 8: Ureter with ureterocele

Traced above the ureter was ending in the lumbar region into a hypoplastic structure with calyces but no evidence of any renal parenchyma. The ureter was filled with a brownish turbid fluid. A cystostomy was done and ureterocele was found extending till bladder neck. The ureterocele was excised along with the ureter and the dysplastic kidney (Figure 9).





**Figure 9:** Excised ureter with calyces and hypoplastic kidney with ureteroceles

A detrusorrhaphy was done and the bladder defect was repaired in two layers. Postoperatively the patient recovered and a cystogram was done which showed a good bladder capacity and function. Histopathologic examination of the specimen revealed small atrophic renal tubules lined by mucin secreting epithelium with no blastemal component suggestive of renal hypoplasia. Ureteric part was lined by transitional epithelial cells.

### 3. Discussion

Ureteroceles are a congenital abnormality characterized by balloon like dilatation of distal ureter near vesicoureteric junction. The embryologic origin of ureteroceles can be best explained as due to the persistence of a thin membrane of tissue between opposing urogenital and ureteric epithelium at the insertion of the ureter into the bladder [1]. Another theory suggests a possible abnormality of collagen or smooth muscle in the wall of ureteroceles [2]. Ureteroceles were classified previously using AAP classification (intravesical or ectopic) [3]. The more commonly used classification system now is the one proposed by Stephens et al (stenotic, sphincteric, sphinctero-stenotic, ectopic, non-obstructive and caecoureteroceles) [4].

The clinical presentation of ureteroceles may be varied, ranging from recurrent urinary tract infections, obstructive voiding symptoms, hematuria, ureteric colic or may be asymptomatic and diagnosed incidentally. Diagnosis of the condition requires a myriad of radiologic investigations, ultrasound being the most commonly used. The typical cobra head sign (dilated ureter surrounded by a thin lucent line) may be seen on intravenous urography. A micturating cystogram is useful investigation to identify the size and type of ureterocoele. It also rules out vesicoureteric reflux [5]. An isotope scan may be invaluable to identify function of kidney in duplex moieties. A cystoscopy may occasionally be required to distinguish the type of ureterocoele [6].

80% of ureteroceles arise in kidneys with a duplex moiety. Females outnumber males in this category. Single system ureteroceles comprise only 20% of the cases with predominantly male distribution [7]. Usually these single system kidneys have good renal function and the ureterocoele is usually intravesical variant [8]. The majority are found in children and usually diagnosed antenatally. Treatment of non-obstructing ureterocoele in single system kidney is usually conservative [9]. Transurethral incision is recommended if the drainage of kidney is affected.

### 4. Conclusion

Usually the kidneys of single system ureterocoele have reasonable function. In our case the kidney was found to be hypoplastic with formed calyces but without functioning renal parenchyma as evidenced on isotope scan. The ureterocoele was also of ectopic type with opening near bladder neck unlike the usual intravesical variant. Extensive literature search could not find any previous reported cases of single system ureteroceles with hypodysplastic non-functioning kidney.

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