

## Cystic Pyeloureteritis: An Uncommon Cause of PUJ Obstruction and Hydronephrosis

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Cystic ureteritis is a disease entity of unknown etiology and symptoms. It is incidentally detected and is difficult to distinguish from other disorders showing filling defects on urographic evaluation. We report a case of PUJ obstruction due to cystic ureteritis that had been diagnosed during ureterorenoscopy in a patient with right-sided hydronephrosis presented with recurrent urinary tract infection. The etiopathogenesis, diagnostic and therapeutic aspects described in the literature are briefly reviewed. We found that endoscopic and histopathological studies are important for confirming the diagnosis along with radiographic findings. Conservative management is advocated for uncomplicated cases. Patient follow up should include urinalysis, urine culture and cytology twice yearly and intravenous urography once a year.

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### Introduction

Cystic ureteritis or cystic pyeloureteritis (CPU) is an unusual benign disease, consisting of the appearance of suburothelial cysts filled with amorphous gelatinous substances that raise the mucosa layer of the urothelium.<sup>1</sup> Most probably a chronic inflammatory process is involved in its pathogenesis. Histopathological examination of pyeloureteritis cystica and cystitis cystica revealed the mechanisms for the cystic formation of the urothelium. Chronic stimulation with inflammation or physical stimulation with crystals or calculi causes the urothelium to form an inflammatory crypt. The crypt is isolated as a result of an adhesive occlusion of the

urothelium at the orifice of the crypt. This crypt is an immature cyst that cannot be clinically detected.<sup>2</sup> A von Brunn's cell nest represents a cut surface of the immature cyst. The inflammatory cyst isolated from the urinary tract, i.e., the immature cyst, gradually grows into a complete cyst, i.e., a clinically visible mature cyst, because of a hydrodynamic flow between the surrounding tissue and neogenetic capillaries, and inflammation. These findings indicate that von Brunn's cell nest, and glandular and cystic formation; occur during development from an inflammatory crypt to an immature cyst and then, a mature cyst.<sup>3</sup>

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### Case Report

A 55-year-old woman from rural Bangladesh, presented with recurrent UTI for long 5 years with occasional right loin to groin pain and fever. She was treated for UTI for several times within this period. Her Urine R/M/E showed numerous pus cells, Urine C/S showed mild growth of *E. Coli*, Serum creatinine 1.2 mg/dl; USG showed moderate hydronephrosis of right kidney with 0.8 cm thick cortex but left kidney was normal and no other remarkable changes. IVU showed right pelvicaliceal dilatation with dye hold up in PCS without visibility of right ureter but normal upper tract on the left side. DTPA renogram showed 19% split function of the right Kidney with poor excretion. Urethroscopy showed no abnormality in bladder and normal ureteric orifice. URS on the right side showed multiple yellowish bubbles arising from the ureteral wall along its whole length extending from very near to vesicoureteral junction to upper third of ureter partially obstructing the lumen of ureter. However most of these bubbles were teased with the grasping forceps. Then left sided ureterorenoscopy showed no pathology there right Kidney explored with the aim of dismembered pyeloplasty. There was nothing inside the renal pelvis except dilatation, no kinking, narrowing at the PUJ or aberrant vessel crossing it.

Opening the PUJ showed multiple bubbles just below the PUJ obstructing ureter lumen completely. After excising the part of renal pelvis and part of the ureter along with it typical A-H pyeloplasty done with D-J stent in situ.

Postoperative period was uneventful, D-J stent removed on 15th postoperative day. Histopathology of the resected renal pelvis and ureter including multiple cysts at PUJ showed nonspecific chronic inflammatory

changes only. Patient was reevaluated for tuberculosis elsewhere, no existing or past history of TB found. Follow up done at 1 month, 3 month, and 9 month and at one year with urine R/M/E, Urine C/S, USG of KUB in every follow up which showed no significant changes except she caught mild UTI at 6 month follow up which have treated accordingly and DTPA renogram at one month and 9 month of operation. Showed 33% and 42% split function of right kidney. Repeat IVU at one year following operative treatment showed well excreting both kidneys with beading in ureter.



Figure 1. Right renal pelvis with part of ureter (resected). Multiple cysts just below pelviureteric junction cause of obstruction.

## Literature Review

The MEDLINE and PubMed data base searched for all English articles and all translated abstract from other language on cystic ureteritis/pyeloureteritis. An additional search was done using the term 'benign lesions of ureter' and causes of Pelvi-ureteric junction obstruction. Recent reviews were checked for additional references.

Ureteritis/Pyeloureteritis cystica characterized by multiple bubbly filling defects in ureter on urography and caused by inflammatory stimuli, are a rare disorder of the ureter. It commonly affects older people. Diagnosis is established by radiological studies.<sup>4</sup> Up to now, no specific treatments except antibiotic therapy are being used to cure this disorder unless one or other complications develop which may require specific treatment. A review of the literature focusing on recently reported cases in the Spanish literature revealed it to be a disease of unknown etiology whose pathogenesis is not well-established, clinical features unspecific, without treatment for the underlying cause, and is a radiologic diagnostic difficulty requiring a differential diagnosis from other filling defects in the urothelium.<sup>5</sup> In one report of a case of pyeloureteritis cystica associated with urinary tract infection and a ureteral stone in a young woman who presented with hematuria and bilateral flank pain.<sup>6</sup> Till now this affection being considered as rare since only about 150 cases have been described in the literature worldwide.<sup>7</sup> It is generally associated with chronic infection and inflammation, and due to its benign nature, treatment must always be conservative and close follow-up is recommended. There are some other rare pathology that may come as differential diagnosis such as Ureteritis follicularis or granularis, uro-tuberculosis, X-ray-negative calculi, UTI with gas-producing germs, Multifocal Transitional Cell Carcinoma,

Vascular ureteral notching, Blood clots, Air bubbles from intervention, Metastases from prostate, stomach, breast (rare), Shistosomiasis, Sloughed papilla. and fibrous ureteral polyps.<sup>8,9</sup> The knowledge of the X-ray diagnostic criteria of the individual diseases is often of life-important significance for making a certain diagnosis for the patient. Another study showed 34 cases of CPU covering the period 1976 to 1994, analyzing the clinical manifestations, diagnostic procedures, differential diagnosis, and evolution. There are no specific symptoms associated with the presence of cysts. The average age of the patients was 59 years (range 30 to 77). Urinary tract infection was detected in 18 (53%). The pyeloureteritis was unilateral in 27 (79%) and bilateral in 7 (21%) of the patients. The location of the cysts was as follows: 1 pyelic (3%); 6 pyeloureteral (18%); and 27 (79%) ureteral.<sup>10</sup> In the pyeloureteritis cystica a bead-like appearance on intravenous pyelogram and retrograde pyelogram as well as in magnetic resonance urography whose resolution depends on the resolution of the associated pathology: infections, lithiasis, and obstruction.

Histopathology of lesions showed: numerous small submucosal epithelial-lined cysts representing cystic degeneration of epithelial cell nests within lamina propria (cell nests of von Brunn) formed by downward proliferation of buds of surface epithelium that have become detached from mucosa.<sup>11</sup> In a case report published in a Spanish literature multiple small submucosal cysts were observed mostly in the pelvic ureter by ureteroscopic examination. Ureteroscopic cold punch biopsy proved ureteritis cystica. The ureteral dilatation improved and filling defects disappeared after the treatment with antibiotics.<sup>12</sup>

A report on a series of 7 new cases of cystic uretero-pyelitis (C.U.P.) presented historical

and pathogenic review of this anatomo-radiologic entity. The conditions for discovery were: renal colic in 4 instances, discovery during surgery in association with a pyeloureteral junction syndrome in one instance, and a hematuria in three instances (1 being accompanied by renal colic).<sup>13</sup> In another case of CPU in association with this condition she had a urinary tract infection due to a coagulase-negative staphylococcus. Following a two-week course of appropriate antibiotic therapy, her urine became sterile and repeat pyelography revealed no abnormality.<sup>14</sup>

### Conclusion

Cystic Ureteritis is a benign and very uncommon condition whose etiology is not well-known. Rarely it may present with one or other complication like the case presented. It is generally associated with chronic infection and inflammation, and may be difficult to distinguish from other filling defects of the urinary tract. Diagnosis is made on the basis of radiologic findings, mainly intravenous urography, endoscopic findings or pathological examination. The disease itself does not require surgical treatment unless it may present with complications.

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