Ureteritis cystica (UC) and xanthogranulomatous pyelonephritis (XP) are rare, benign conditions that uncommonly appear together. The first one has not well established etiology yet and can affect ureter, renal pelvis and bladder. The second one is a chronic renal infection resulting in severe kidney deterioration and destruction. Both of these entities are usually unilateral. They both cause wide spectrum of symptoms so the diagnose can be difficult to establish.

We report the case of a 68 year old male with a history of intermittent macroscopic hematuria, left flank pain, recurrent urinary tract infection and calculosis of left kidney. We describe the imaging and endoscopic investigations leading to the diagnosis of ureteritis cystica as well as the treatment options.

Key words: Ureteritis cystica, Xanthogranulomatous pyelonephritis, Kidney

INTRODUCTION

Ureteritis cystica (UC) is a rare, benign condition that usually affects the ureter but can reach the renal pelvis as well as the bladder. It is first described in 1761 by Morgagni\textsuperscript{1,2}, who referred two cases and after him confirmed by Johnson in 1816 and Rayer in 1837 and Rokitansky in 1861\textsuperscript{1,2}.

This condition differs in its appearance; it is usually asymptomatic but infection and obstruction caused by the cysts may present symptoms of hematuria, chronic pyelonephritis or/and renal calculi. In asymptomatic patients ureteritis cystica is detected accidentally with intravenous urography, retrograde ureterography and ureteroscopy during resolving other underlying pathologic changes. Ureteritis cystica is usually suspected when defects of filling are seen along the ureter in contrasted images of the urinary tract. It is not uncommon for this condition to presents with acute flank pain.

Xanthogranulomatous pyelonephritis is a chronic renal infection associated with obstructive uropathy due to nephrolithiasis.\textsuperscript{3} Chronic obstruction by calculi and infection are major pathogenetic factors in this disease. The kidney becomes enlarged and over time it is destroyed starting with pelvis and calyces followed by parenchymal deterioration. Patient with XP has symptoms of persistent bacteriuria (49%), flank pain (69%), fever and chills (69%)\textsuperscript{3}.

The case described here illustrates this rare form of presentation. We describe the imaging and endoscopic investigations leading to the diagnosis of ureteritis cystica as well as the treatment of this pathological condition.

CASE REPORT

A 68-year-old man was admitted complaining intermittent macroscopic hematuria, flank pain and recurrent urinary infection. His medical history presented diabetes mellitus, hypertension and coronary insufficiency followed by stent of right coronary artery. Patient had an episode of spontaneous calculi elimination three years before admission. Blood levels of urea, creatinine, electrolytes and red blood cells count were in normal ranges. Leukocytes were raised to 12x10\textsuperscript{9} and body temperature varied between 37,2 and 38,4°C. A urinalysis showed a pH of 6, was positive for haemoglobin and leukocyte esterase (++). The sediment showed 55.4 epithelial cells/\mu L and 1460 bacteria/\mu L. Voided urinary cytology was negative for malignant cells. DTPA renal scan showed renal function R:L=89:11%.

After administering fluids and I.V antibiotics, abdominal ultrasound was performed as a first step in establishing diagnosis. It showed calculi in middle and...
upper calices of the left kidney which had slightly hypotrophic appearance.

Intravenous urography that followed, confirmed stones in upper and middle calices of left kidney. (Figure 1)

Filling defects were obvious finding along the renal pelvis, ureteropelvic junction and ureter.

Computerized tomography (CT) was performed and stone disease of left kidney was confirmed. CT scan didn’t show pathological finding in pelvis and ureter. (Figure 2).

Cistocopy showed normal bladder mucosa but ureteroscopy of left ureter was unsuccessful because of bleeding that occured.

Considering all above mentioned findings and the patient condition, an indication for open nephroureterectomy was made. Before left nephroureterectomy was done “ex tempore” biopsy was performed. Histopathological finding presented tubular thyroidization, interstitial growth of fibrous tissue and chronic interstitial inflammation. Pelvis and ureter presented intact surface of mucosa with islands and invaginated glandular structures originated from uroepithelium in submucous space without atipic epithel cells.

Removed specimen was kidney; it weight 95 grams with dimension 10x5 cm, 22cm long ureter with bladder cuff. Seven stones were removed from dissected kidney; proximal and middle calices were dilated and renal parenchyma was 5-10 mm thin. Along the pelvis and the ureter multiple cysts with diameter up to 4 mm were found. Regional lymphoglandules showed chronic inflammation. (Figure 3, 4)

**DISCUSSION**

Ureteritis cystica is defined as the cystic transformation of the epithelial nest of Brunn. In 1876 Litten offered the first microscopic description of these cysts, but without nests. The cysts are well defined and containing clear fluid with sizes between 1 and 10 mm and flattened epithelial walls probably as the result of irritation in nonspecific chronic inflammations. Pierrard described ureteritis cystica as a benign inflammatory disease of the urinary tract, resulting from chronic inflammation of the urothelium. Possible etiological factors include bilharziasis, vitamin A excess and increased immunoglobulin A. Ureteritis cystica is a rare condition that predominantly affects adult females, but it is also reported to occur in men and children. It is usually unilateral, but bilateral cases have been described as well. The cysts can be found at any level of the urothelium. When present in the bladder they are referred to as cystitis cystica. The lesions are benign with low potential for degeneration, although very occasionally it has been associated with bladder carcinoma or renal carcinoma. Macroscopically it manifests with multiple cysts that make filling defects which are difficult to differ from multiple tumors of renal pelvis and the ureter, tuberculosis, lesions caused by gas-forming microorganisms and radiolucent calculi with imagining techniques. The clinical presentation is variable, an it usually depend on associate pathologic conditions like urinary tract infections (82%), lithiasis (53%) or haematuria (52%).

![Figure 1: Excretory Pyelography of the Patient T.S (With Permission Obtained)](image1)

![Figure 2: Cystoscopy Showing Normal Bladder Mucosa but Ureteroscopy of Left Ureter was Unsuccessful Because of Bleeding That Occurred.](image2)
The most commonly used imaging techniques in establishing diagnosis of urinary tract are excretory urography and retrograde pyelography which show filling defects in affected part. In cases where the diagnosis is uncertain, as was the case in our patient, CT scan and MRU can be used to better define the nature of the lesions, their extent, or the presence of other abnormalities. Other lesions that can have a similar appearance include radiolucent stones, clots, veins, air.

Ureteroscopy gives the most certain insight in this condition because it enables the direct visualization of the cystic formations and the opportunity to biopsy them in order to realize an pathohistological analysis.

Xanthogranulomatous pyelonephritis is a rare chronic infectious disease of the kidney associated with nephrolithiasis. Review of the literature shows Proteus to be the most common microorganism but E.colli is very common as well. This condition is almost always unilateral and 50-80% of the patients manifest it with triad of unilateral renal enlargement with little or no function and a calculus that very commonly can be staghorn. Because of its unilateral appearance the values of serum degradation products remain normal. Xanthogranulomatous pyelonephritis can imitate every other inflammatory kidney disease as well as RCC on radiographic examination. CT is most useful in diagnosing this condition and radionuclide scan using 99m Tc-DMSA is used to confirm and quantify the differential lack of function of the affected kidney.

When the renal function is proven to be deteriorated nephrectomy is the only option in treating this pathologic condition.
The suggested treatment for ureteritis cystica on the opposite side is conservative by eliminating the process which is causing the inflammation (infection, lithiasis).

But in cases where UC is associated with renal destruction due to xanthogranulomatous pyelonephritis surgery is still a choice of treatment.

**CONCLUSION**

Ureteritis cystica is a benign condition often associated to other urological diseases. The basic diagnostic procedures are imaging studies complemented with ureteroscopy and biopsy. Treatment of the underlying cause and watchful observation are the mainstays of management. The case presented is unique because its presentation and treatment differ because of the combined pathological conditions that affected both the kidney and the ureter.

**SAŽETAK**

Ureteritis cystica (UC) i ksantogranulomatozni pijelonefritis (KSP) su retka, benigna oboljenja urinarnog trakta koje se obično ne javljaju zajedno. UC je još uvek sa nedovoljno poznatom etiologijom i može da zahvati ureter, bubrežnu karlicu i bešiku. KSP je hronična bubrežna infekcija koja dovodi do teškog oštećenja i destrukcije bubrega. Oba entiteta su obično jednostrana. Dovode do širokog spektra simptoma i dijagnoza se može teško utvrditi. Prikazujemo slučaj muškaraca starog 68 godina sa istorijom intermitentne makroskopske hematurije, bolovima u levom boku, rekurentnim infekcijama urinarnog trakta i kalkulozom levog bubrega. Opisane su imaging i endoskopske istrage koje vode ka dijagnozi UC kao i mogućnosti lečenja.

Ključne reči: ureteritis cystica, ksantogranulomatozni pijelonefritis, bubreg

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