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A CASE OF ISOLATED AUTOIMMUNE URETERITIS IN A 25-YEAR-OLD MALE

Ionut Codrin Gheorghe Hogea[1], James Albert Geroge Littlewood[1], Snehal Patel[2], Noshaba Naz[1], Khaled Abdulnabi[1]

[1] Nephrology Department, Wirral University Teaching Hospital, Upton, UK
[2] Urology Department, Wirral University Teaching Hospital, Upton, UK

Corresponding author's e-mail: codrin.hogea@gmail.com

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ABSTRACT

Background: Ureteritis is an inflammatory condition of the ureters, most commonly caused by infection, often associated with cystitis or pyelonephritis. Non-infectious causes, including autoimmune diseases, are rare and pose significant diagnostic challenges. Autoimmune ureteritis is an exceedingly rare entity with limited literature guidance, often diagnosed by exclusion.

Case description: We report the complex six-year clinical course of a 25-year-old male who initially presented with right flank pain and macroscopic haematuria. Early investigations including imaging and cystoscopy were unremarkable, leading to a provisional diagnosis of loin pain haematuria syndrome. Two years later, worsening symptoms prompted repeat imaging, which revealed bilateral ureteric and renal pelvic thickening. Multiple investigations, including extensive infectious screening, PET-CT and serial ureteroscopies with biopsies, failed to identify a definitive cause. Histopathology remained non-diagnostic; empirical antibiotic therapy was ineffective. A trial of corticosteroids resulted in significant symptom improvement and radiological resolution of inflammation, strongly suggesting an autoimmune aetiology. Immunosuppressive therapy with mycophenolate mofetil, later switched to azathioprine, maintained remission. The patient remains in remission, having finished a two-year course of azathioprine.

Conclusion: This case highlights the diagnostic complexity of autoimmune ureteritis, particularly in the absence of systemic autoimmune disease. The diagnosis was made by exclusion after thorough investigation ruled out infection, malignancy and systemic autoimmune conditions. The patient's positive response to immunosuppression supports an immune-mediated pathogenesis. Isolated autoimmune ureteritis should be considered in patients with persistent ureteric inflammation, and negative infectious and malignant work-up. Early recognition and immunosuppressive therapy may prevent long-term structural damage and preserve renal function.

KEYWORDS

Immunosuppressive therapy, ureteroscopy, autoimmune ureteritis

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LEARNING POINTS

- This case demonstrates the diagnostic complexity of isolated autoimmune ureteritis, highlighting the value of excluding the common differentials.
- It highlights the limitations of diagnostic tools in ureteric disease, reinforcing the role of clinical judgement and treatment trials when histology is inconclusive.
- It illustrates the role of immunosuppressive therapy in rare inflammatory conditions, not only as an effective treatment but also as a diagnostic tool.

INTRODUCTION

Ureteritis is an uncommon condition characterised by inflammation of the ureters. The most frequent aetiology is infectious, often associated with cystitis or pyelonephritis[1]. The predominant causative organisms include *Escherichia coli*, *Staphylococcus spp.*, *Streptococcus spp.*, *Enterococcus*, *Proteus*, and *Pseudomonas aeruginosa*[2].

Aseptic inflammation of the upper urinary tract, including the renal pelvis and ureters, has also been described in conditions such as inflammatory pseudo tumour of the ureter, idiopathic segmental ureteritis, idiopathic retroperitoneal fibrosis and eosinophilic ureteritis[3]. Additionally, non-infectious ureteritis may occur in individuals with pre-existing autoimmune diseases, including rheumatoid arthritis and systemic lupus erythematosus[4,5].

CASE DESCRIPTION

We present the six-year clinical course of a 25-year-old male who presented with severe right flank and loin pain, accompanied by small clot haematuria. His care involved a multidisciplinary approach, led primarily by nephrology with regular input from the urology team.

The patient had a background of hay fever, high body mass index (BMI) and previous inguinal hernia repair. On initial presentation in 2018 his renal function was normal. A renal angiogram, cystoscopy and right sided ureteroscopy were unremarkable. He was subsequently diagnosed with loin pain haematuria syndrome and discharged home.

The patient re-presented two years later with worsening symptoms (intermittent macroscopic haematuria with clots and ongoing right-sided flank pain) requiring strong opioid analgesia. He reported regular visits to Iran and Iraq, as well as recent travel to Dubai and Spain, but there were no rural visits of exposure to freshwater.

His renal function remained normal. A renal biopsy was performed, initially suggesting IgA nephropathy, but ultimately proving to be inconclusive. A repeat computed tomography (CT) urogram demonstrated abnormal bilateral thickening of the ureters and renal pelvises (more pronounced on the right). This new finding was more suggestive of an infective or inflammatory aetiology and not in keeping with the previous diagnosis of loin pain haematuria syndrome. The possibility of a colovesical fistula was also ruled out.

The patient then underwent a cystoscopy with right-sided ureteroscopy for direct visualisation of the urinary tract. This corroborated the findings of the cross-sectional imagining, revealing that the right upper ureter was inflamed, and

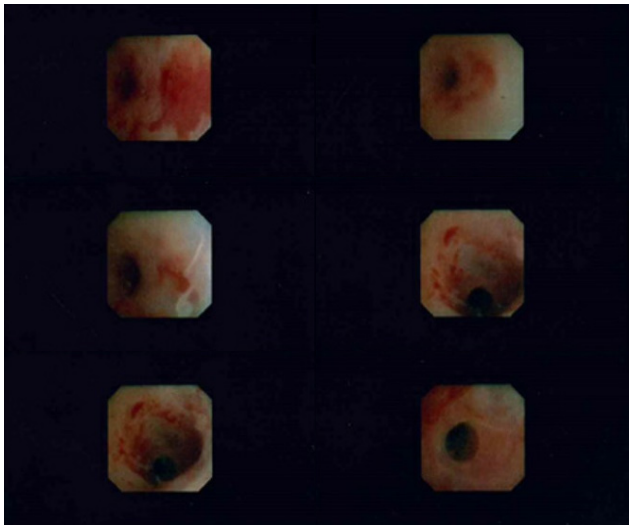


Figure 1. Ureteroscopy showing right-sided ureteral inflammation.

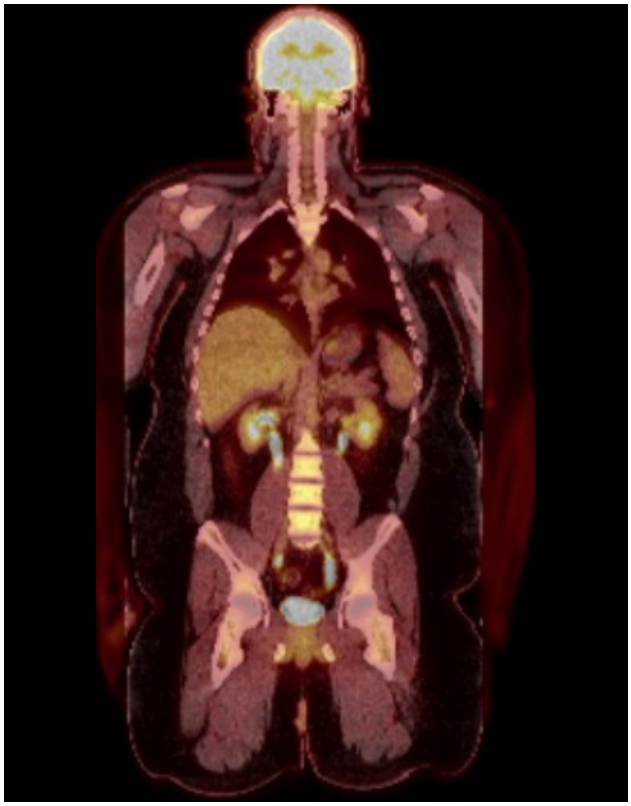


Figure 2. PET scan showing no abnormal FDG activity in the ureters.

identifying an abundance of white flesh particles in the bladder which raised the suspicion of parasitic casts *(Fig. 1)*.

As a result of this finding and taking into account the patient's travel history, an extensive infectious screening was performed. Urinalysis revealed high numbers of leukocytes and increased levels of oxalate and citrate, but the infectious screening (including cytomegalovirus, schistosomiasis, filariasis, hydatid disease and tuberculosis) was negative. The ureteral biopsy performed at that point showed no evidence of inflammation or parasitic presence.

A renogram showed minor anatomical abnormalities with a slightly delayed peak uptake in the right kidney (left: 46%, right: 54%). A PET-CT scan revealed no abnormal fluorodeoxyglucose (FDG) activity in the renal parenchyma, prostate, seminal vesicles or testes *(Fig. 2)*.

A six-week empirical course of trimethoprim, allopurinol and bicarbonate was initiated to address occult infection and alkalinise the urine but led to no clinical improvement.

Approximately four months later symptoms worsened, raising the suspicion of papillary necrosis. A repeat CT urogram, however, showed stable ureteric appearances and ruled out this diagnosis. Repeat ureteroscopy and pyelography confirmed persistent inflammation in the right upper ureter, with similar findings to the previous procedure. Biopsies were again non-diagnostic, as tissue samples did not survive processing. The case was discussed at the benign uro-radiology multidisciplinary team, where it was suggested that the inflammation appeared periureteral rather than intraureteral, making retroperitoneal fibrosis or autoimmune ureteritis possible causes. IgG4 serology was negative, making the former less likely. Given these findings, the patient was commenced on a trial of corticosteroids and reported significant symptomatic relief over the next three months. He was transitioned to mycophenolate mofetil with a plan to taper the steroids.

A CT urogram at this time showed significant radiological improvement, with resolution of the left-sided changes and only subtle soft tissue thickening along the right ureter *(Fig. 3)*.

Approximately six months later, the patient re-presented with recurrent symptoms, likely indicative of treatment failure. A repeat CT urogram showed worsening thickening of the right ureter. Ureteroscopy and pyelography were once again performed, and this time showed structural changes in the upper ureter *(Fig. 4)*.

Endoscopic examination of the ureter revealed the presence of a band of white, pale tissue akin to a benign structure, as opposed to the inflammatory changes visualised on previous procedures.

A biopsy was once again non-diagnostic, showing only a few detached urothelial cells with reactive changes *(Fig. 5)*.

As the clinical picture was indicative of a relapse, the immunosuppressant agents were adjusted: mycophenolate mofetil was stopped and replaced with azathioprine. A low dose of maintenance steroids was continued.

Due to the development of structural ureteric changes, a

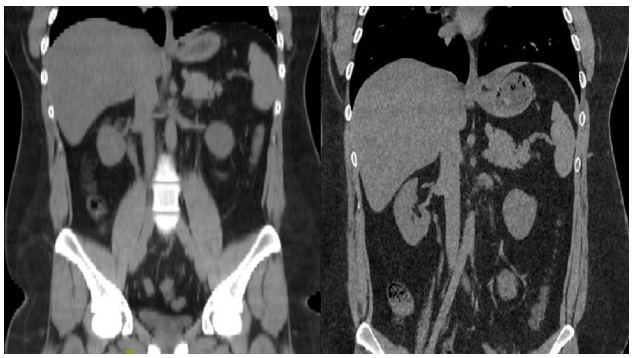


Figure 3. Interval radiologic improvement in ureteric appearances following a three-month course of steroids and mycophenolate mofetil therapy.



Figure 4. Pyelography showing ureteric stricture.

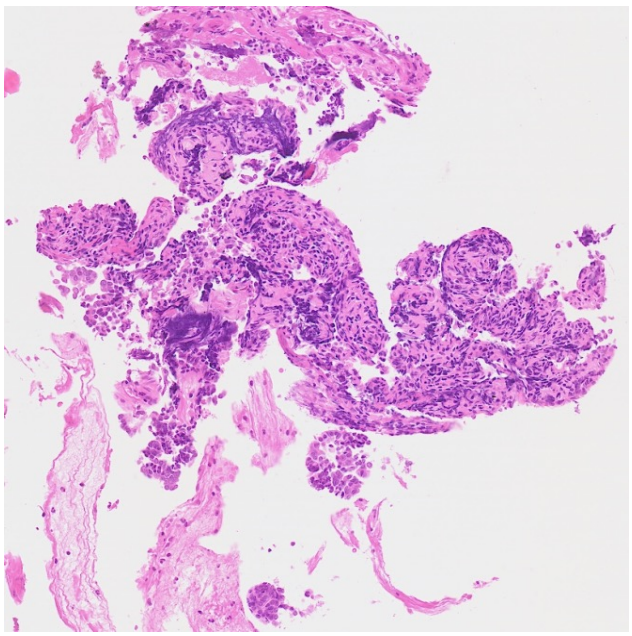


Figure 5. Ureteral biopsy showing detached urothelial cells with reactive changes. There is no nuclear enlargement, hypercromasia or mitosis. The underlying fibro-vascular stroma contains scattered chronic inflammatory cells; there is no evidence of carcinoma in situ or malignancy.

renogram was performed to assess the dynamic drainage of the kidneys. It demonstrated stable renal function with interval improvement in drainage characteristics compared to a 2021 scan.

The patient remained under regular nephrology follow-up and reported sustained symptomatic improvement after the initiation of azathioprine. Immunosuppressive therapy has since been gradually tapered with no evidence of disease recurrence. He was formally discharged by the urology team in November 2023 and currently remains well and asymptomatic, attending biannual nephrology reviews.

DISCUSSION

Autoimmune ureteritis is an exceedingly rare diagnosis, typically considered a diagnosis of exclusion. Prior to suspecting an autoimmune aetiology, it is essential to rule out more common causes including active parasitic infections, genitourinary malignancies and urinary calculi. The majority of ureteritis cases are of infectious origin, often caused by organisms commonly associated with urinary tract infections, such as *Escherichia coli*, *Staphylococcus spp.*, *Streptococcus spp.*, *Enterococcus*, *Proteus* and *Pseudomonas aeruginosa*[2].

Parasitic infections, although uncommon in the genitourinary tract, are typically chronic in nature. The most frequently implicated parasites include *Schistosoma*, *Echinococcus* and *Wuchereria bancrofti*[6].

In our case, an extensive infectious screening was performed and returned negative, with the exception of two isolated instances of positive urine cultures, consistent with acute rather than chronic urinary tract infection. A six-week empirical course of trimethoprim yielded no symptomatic improvement, further suggesting that chronic infection was unlikely to be the underlying cause of the symptoms.

Cross-sectional imaging excluded the presence of urinary tract calculi or malignancy. Furthermore, PET imaging demonstrated normal FDG uptake within the renal parenchyma. Endoscopic biopsy is critical in the diagnosis, monitoring and management of upper ureter and renal pelvis urothelial lesions[7]. In our case, initial endoscopic visualisation revealed non-specific inflammatory changes, which progressed to structural abnormalities over time. However, multiple biopsies either showed only non-specific inflammation or were non-diagnostic, precluding a definitive histological diagnosis. The initial ureteral biopsy was normal and did not show any inflammatory changes or parasitic presence. The second biopsy was uninterpretable as no tissue survived processing. The third biopsy, performed three years after the first one, only showed chronic reactive inflammatory changes.

This is not surprising, as proximal ureteral and renal pelvis biopsies are challenging to obtain not only due to anatomical reasons, but also due to technical difficulties of ureteral tissue sampling[7]. As such, most samples are insufficient, superficial or lacking depth, making slide interpretation difficult. Moreover, the presence of artifacts or distorted architecture makes the morphology unreliable. This leads to almost 1 in 4 cases of ureteral biopsies not yielding a definitive diagnosis[7].

Following the exclusion of infectious, malignant and structural causes, an immune-mediated aetiology was considered. Ureteritis associated with immunological disorders is extremely rare, with most reported cases involving small-vessel vasculitis or immunoglobulin G4-related disease (IgG4-RD)[4]. Diagnosis of IgG4-RD typically relies on elevated serum IgG4 levels (≥1.35 g/l) together with histopathological findings of lymphoplasmacytic infiltration, abundant IgG4-positive plasma cells and fibrosis[8]. In this case, serum IgG4 levels were within normal limits, and ureteral biopsy findings were non-diagnostic, thereby making IgG4-RD unlikely. Similarly, complement levels were normal and dsDNA and anti-CCP antibodies were negative, rendering lupus and rheumatoid arthritis unlikely differentials.

With all other causes excluded an autoimmune process was suspected, and the patient was commenced on immunosuppressive therapy. The initiation of corticosteroids led to a marked clinical improvement, supporting the hypothesis of an autoimmune aetiology. This was further corroborated by interval imaging, which demonstrated structural improvement following treatment.

CONCLUSION

Evidence on the treatment of autoimmune ureteritis is sparse, largely limited to case reports or instances where ureteritis occurred in the context of a known autoimmune condition. A review of the literature suggests that immunosuppressive therapy, often in combination with ureteral stenting, is the cornerstone of management. Corticosteroids have shown efficacy in patients with rheumatoid arthritis-associated ureteritis[4], and prednisolone has been successfully used to treat immune checkpoint inhibitor-induced ureteritis[9]. In systemic lupus erythematosus-related ureteritis, a combination of steroids and mycophenolate mofetil has been effective[5]. Cyclophosphamide, alongside corticosteroids, has also been employed in cases of lupus-related cystitis complicated by hydroureteronephrosis[10].

This case is notable for its diagnostic complexity, the final diagnosis being isolated as autoimmune ureteritis. The patient's favourable response to immunosuppressive therapy strongly supports an underlying autoimmune mechanism. He has remained in clinical remission and is now off immunosuppression, having completed a two-year weaning course of azathioprine.

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