Journal of Pediatric Urology (2010) 6, 538—542

Journal of Pediatric urology

REVIEW ARTICLE

Stenosing ureteritis in a 7-year-old boy with Henoch—Schönlein purpura nephritis: A case report and review of the literature☆

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Received 4 November 2009; accepted 1 March 2010
Available online 10 April 2010

KEYWORDS
Henoch—Schönlein purpura;
Stenosis;
Renal pelvis;
Ureter;
Nephritis;
Obstruction;
Ureterocalycostomy

Abstract *Introduction:* Urinary tract obstruction resulting from Henoch—Schönlein purpura (HSP) is an extremely rare urologic entity. If the genitourinary system is involved, it is primarily in the form of a focal proliferative glomerulonephritis. Stenosing disease has received little attention in the literature and treatment options are limited. Despite early intervention renal loss may be inevitable.
Case report: A 7-year-old African American male presented with renal failure secondary to bilateral sclerosing ureteritis 1 month after initial presentation with HSP. There was significant disease progression and he required multiple procedures, ultimately bilateral ureterocalycostomies and a left nephrectomy.
Discussion: HSP is an immune-mediated necrotizing vasculitis. It can affect any organ system; however, in the genitourinary system, focal proliferative glomerulonephritis is a common manifestation, occurring in 20—90% of cases [8]. Extrarenal manifestations are rare.
Conclusion: Ureteritis and obstruction may be late occurrences, but should be considered in all patients presenting with a history of HSP and new-onset flank pain, acute renal failure, or

☆ Support and financial declaration: There was no research support for this paper. In addition, none of the listed authors has any financial conflicts of interest associated with this paper.

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doi:10.1016/j.jpurol.2010.03.001

anuria. Families and patients should be counseled that renal impairment may be a consequence of stenosing ureteritis. Management of these patients can be complicated but surgical correction must be considered in the treatment algorithm once the disease has stabilized.

Introduction

Henoch—Schönlein purpura (HSP) is a common systemic vasculitic condition of which the majority of cases occur in the pediatric population. It is an immune-mediated disorder characterized by tissue deposition of immunoglobulin A (IgA) complexes. Although the renal injury may in part be mediated by immunoglobulin G (IgG) autoantibodies directed against mesangial cell antigens, it is the predominance of IgA immunofluorescence that is the hallmark of HSP nephritis. The majority of cases are self-limited, and common clinical features include a non-thrombocytopenic purpuric rash, arthritis, abdominal pain, and nephritis [1]. Approximately 20—50% of cases of HSP are preceded by an upper respiratory tract infection, most commonly the result of a streptococcal infection [2]. Genitourinary complications of HSP are rare, with scrotal symptoms of swelling and pain, or epididymitis being the most common in 13—20% of male patients [3,4]. Ureteritis, either hemorrhagic or sclerosing, is an extremely rare complication, but often requires surgical intervention. We report the case of a 7-year-old male with severe acute renal failure (ARF) secondary to bilateral sclerosing ureteritis 1 month after initial presentation with HSP. In addition to the medical management he received, he also required complex surgical intervention to reconstruct his urinary system.

Case report

A 7-year-old African American male presented to our emergency center with a 1-week history of pharyngitis, tonsillar enlargement, ulcers on the buccal surface, low-grade fever, and arthritis involving the wrists, ankles and right elbow. His blood pressure was elevated (179/92) and his laboratory evaluation revealed 3+ proteinuria and 3+ hematuria, serum albumin 2.6, hyponatremia of 130, BUN/Cr (blood urea nitrogen to creatinine ratio) of 16/0.6, leukocyte count of 12.9 K, anemia with hemoglobin 9.9, thrombocytosis of 530 K, C-reactive protein 5.3, erythrocyte sedimentation rate 71, normal C3 of 116, and low C4 of 5 (range 14—36). Antinuclear antibody was reactive and a rapid streptococcal throat swab was positive. Blood, urine, and cerebrospinal fluid cultures grew no organisms. Renal ultrasound demonstrated normal, symmetric kidneys (right 8.5 cm and left 9 cm) with no hydronephrosis and only a mild increase in echogenicity bilaterally (Fig. 1). The patient was admitted and treated with intravenous (IV) vancomycin and cefotaxime for possible sepsis. Five days into the illness, the patient developed a violaceous rash on his legs, ankles, and feet.

The arthritis, glomerulonephritis, and purpuric rash were consistent with a diagnosis of HSP, but the low C4, severe hypoalbuminemia, antinuclear antibody reactivity, and anemia were not. A renal biopsy was therefore performed which demonstrated HSP nephritis (Fig. 2). The patient was treated with IV furosemide for diuresis, with improvement in his blood pressure. After a few more days the patient was stable and discharged home.

However, he returned to the emergency center 1.5 months after initial presentation with vomiting for 2 weeks and anuria for 18 h. He was otherwise without symptoms. Laboratory evaluation demonstrated ARF with serum Cr 16.2 and BUN 106. His serum potassium was elevated at 6.6 but all other electrolytes were within normal limits and his hypoalbuminemia, leukocyte count, anemia, and thrombocytosis were all improved. Despite fluid boluses to correct dehydration and urethral catheter placement, he continued to be anuric. Emergency renal biopsy was

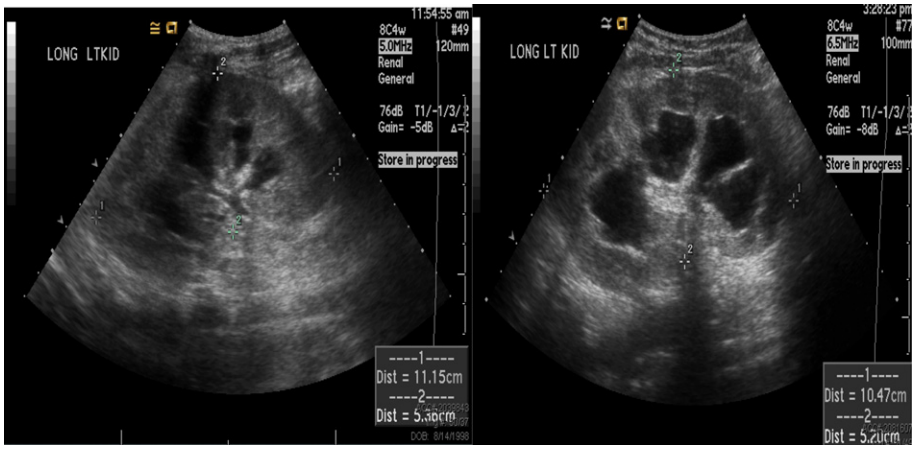

Figure 1 Renal ultrasound — left kidney early in disease (image on left) and with disease progression and worsening caliectasis (image on right).