

## Bilateral ureteral stenosis as a complication of Henoch–Schönlein vasculitis

Chieko Ihoriya · Yoshitaka Morita · Takehiko Tokura ·  
Kengo Kidokoro · Norio Komai · Tamaki Sasaki ·  
Naoki Kashihara

Received: 16 January 2008 / Accepted: 25 February 2008 / Published online: 22 April 2008  
© Japan College of Rheumatology 2008

**Keywords** Ureteral stenosis ·  
Henoch–Schönlein purpura · Vasculitis

We reported previously on a case of a young adult with dermatomyositis who developed bilateral ureteral stenosis [1]. Although rare, ureteral involvement can be present in various forms of systemic small-vessel vasculitis [2]. However, this complication can be overlooked by rheumatologists. Indeed, there are only a few reports of this complication in the rheumatology literature. Here we report on another case of bilateral ureteral stenosis in a patient with Henoch–Schönlein vasculitis.

A 66-year-old male was referred to our clinic in January 2007 for aggravation of renal failure. He was admitted to another hospital in December 2006 for macrohematuria. He reported that he hadn't had any abnormal signs or symptoms during the last few months. Physical examination in that hospital revealed normal chest and abdomen and no skin rash or purpura. Laboratory findings included leukocyte count of 17,460/ $\mu$ l, C-reactive protein (CRP) 10.1 mg/dl, serum creatinine 1.95 mg/dl, and blood urea nitrogen of 18 mg/dl. A computed tomographic scan demonstrated bilateral hydronephrosis. Intravenous pyelography revealed complete obstruction in the left lower ureter, and severe stenosis in the right lower ureter. Intracystic endoscopy showed no abnormalities in the bladder mucosa. Due to the evident renal dysfunction, a stent was inserted into the right

stenotic ureter. Percutaneous nephrostomy was performed for hydronephrosis of the left kidney (Fig. 1). However, serum creatinine increased further, and he was referred to our clinic for further management.

Laboratory tests at our clinic showed a leukocyte count of 9,680/ $\mu$ l, an erythrocyte count of  $282 \times 10^4$ / $\mu$ l, a platelet count of  $27.5 \times 10^4$ / $\mu$ l, hemoglobin 8.1 g/dl, CRP 7.30 mg/dl (normal <0.30), serum creatinine 5.32 mg/dl, and blood urea nitrogen 28 mg/dl. Serological tests were negative for cryoglobulin, antinuclear antibody, and antineutrophil cytoplasmic antibodies. Serum concentrations of complements (C3 and C4) were normal. The levels of both IgG and IgM were normal, but IgA levels were increased to 998.3 mg/dl. Prothrombin time (PT) and activated partial thromboplastin time (APTT) were normal. Urinalysis showed numerous red blood cells with granular and white cell casts under high-power field. Urine protein was positive at 2.06 g/day. Renal biopsy was performed and the histology showed necrotizing glomerulonephritis with cellular crescents (Fig. 2). Five glomeruli with global sclerosis were noted. Cellular crescents were found in four out of the remaining nine glomeruli. Immunofluorescence histochemistry showed fine granular deposition of IgA in the paramesangial areas. Immune-complex deposits were also confirmed by electron microscopy. The patient was diagnosed as having Henoch–Schönlein vasculitis. He was treated with 30 mg/day prednisolone, which resulted in a rapid normalization of CRP and improvement in serum creatinine levels.

However, two weeks after starting the steroid therapy, the patient developed massive melena. Erythrocyte count and hemoglobin were decreased at  $173 \times 10^4$ / $\mu$ l and 5.5 g/dl, respectively. Platelet count was  $15.4 \times 10^4$ / $\mu$ l. PT and APTT were not prolonged at 10.0 and 21.0 s,

C. Ihoriya · Y. Morita (✉) · T. Tokura · K. Kidokoro ·  
N. Komai · T. Sasaki · N. Kashihara  
Division of Nephrology and Rheumatology,  
Department of Internal Medicine, Kawasaki Medical School,  
577 Matsushima, Kurashiki, Okayama 701-0192, Japan  
e-mail: morita@med.kawasaki-m.ac.jp

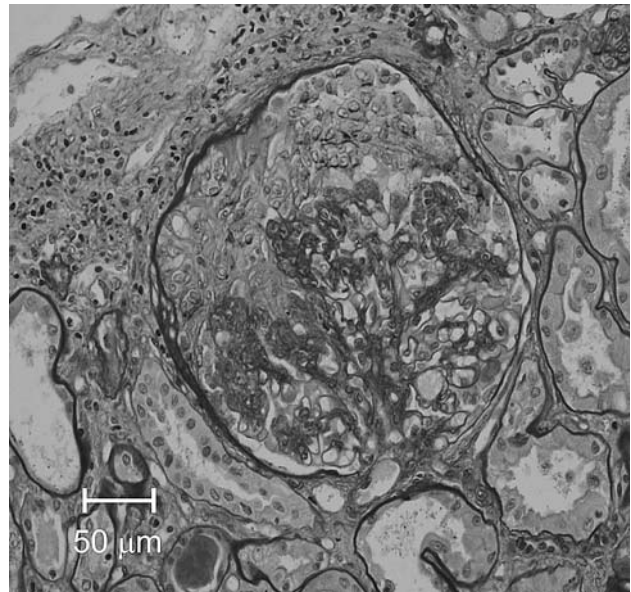


**Fig. 1** Percutaneous pyelography demonstrating stenosis of the left lower ureter (*arrow*). Note the left hydronephrosis with a dilated ureter above the stenosis. A stent is inserted into the right ureter

respectively. Serum CRP level was not elevated at 0.11 mg/dl. Serum creatinine level was 2.62 mg/dl. Since the source of the bleeding could not be localized by upper or lower intestinal endoscopy, it was considered to be the small intestine. Therefore, it was suspected that intestinal bleeding was associated with Henoch–Schönlein vasculitis. Accordingly, cyclophosphamide (25–50 mg/day) was added to the steroid therapy to control the activity of the systemic disease. The melena subsided with conservative treatment.

Follow-up percutaneous pyelography performed in February demonstrated partial resolution of the left ureteral obstruction. The catheter for left nephrostomy was removed and a stent was inserted into the stenotic ureter. In March, he was discharged from the hospital with bilateral ureteral stents. He remains clinically well and is being treated with 5 mg/day prednisolone at the time of writing of this report.

Although it is extremely uncommon, the ureter may be involved in Henoch–Schönlein purpura. A literature search of the PubMed database identified nine documented cases of patients with Henoch–Schönlein purpura who developed ureteral stenosis (six of them in non-English publications



**Fig. 2** Renal biopsy specimen. Necrotizing glomerulonephritis with cellular crescents (PAS stain,  $\times 200$ )

with English abstracts) [3–11]. The previous cases occurred at the ages of 6 to 14. Purpura was not observed in our patient. However, clinical and laboratory findings, such as necrotizing glomerulonephritis with cellular crescents and mesangial IgA deposition, intestinal bleeding and systemic inflammation, supported the diagnosis of Henoch–Schönlein vasculitis in our case.

Ureteral involvement may lead to serious complications in patients with systemic small-vessel vasculopathy. Hematuria in patients with vasculitis suggests renal involvement, but the possibility of ureteral stenosis should also be considered. Early diagnosis and treatment are crucial for long-term prognosis. Computed tomography or renal ultrasonography may help identify the presence of ureteral stenosis. The present case emphasizes the importance of recognizing this complication in systemic vasculopathy including Henoch–Schönlein purpura.

## References

1. Morita Y, Sakuta T, Nagasu H, Kuwabara A, Tokuoka Y, Teshigawara S et al. Bilateral ureteral stenosis and duodenal perforation in dermatomyositis. *Mod Rheumatol*. 2007;17:54–6.
2. Baskin L, Mee S, Matthey M, Carroll PR. Ureteral obstruction caused by vasculitis. *J Urol*. 1989;141:933–5.
3. Pfister C, Liard-Zmuda A, Dacher J, Dubois D, Grise P, Mitrofanoff P. Total bilateral ureteral replacement for stenosing ureteritis in Henoch–Schönlein purpura. *Eur Urol*. 2000;38:96–9.
4. Smet MH, Marchal G, Oyen R, Breysem L. Stenosing hemorrhagic ureteritis in a child with Henoch–Schönlein purpura: CT appearance. *J Comput Assist Tomogr*. 1991;15:326–8.
5. Kher KK, Sheth KJ, Makker SP. Stenosing ureteritis in Henoch–Schönlein purpura. *J Urol*. 1983;129:1040–2.

6. Maherzi A, Kaabar N, Boussetta K, Salem M, Hammou A, Chaouachi B et al. Bilateral stenosing ureteritis in Henoch–Schönlein purpura. *Arch Pediatr*. 1997;4:36–9 (In French).
7. Bosio M, Ravelli A, Ruperto N, Migliori C, Perotti F, Scotta MS, Martini A. The Schönlein–Henoch syndrome with severe multi-systemic involvement. *Minerva Pediatr*. 1993;45:197–201 (In Italian).
8. Kurunczi S, Görke WD, Sperling G. Stenosing ureteritis and factor XIII deficiency in anaphylactoid purpura. *Z Kinderchir*. 1990;45:314–6 (In German).
9. García Mérida M, Quiñonero Díaz A, Bueno Fernández A, Galiano Duro E, López Pérez GA, Miguélez Lago C. Pyeloureteral stenosis secondary to Schoenlein–Henoch purpura. *Actas Urol Esp*. 1989;13:118–20 (In Spanish).
10. Grasso E, Bianchi GP, Candeo G, Sticca M. A case of stenosing ureteritis in Schoenlein–Henoch purpura. *Pediatr Med Chir*. 1988;10:319–21 (In Italian).
11. Hayat P, Sonsino E, Bompard Y, Stora-Castaing N, Weisgerber G. Ischaemic stenosis of the ureter during Henoch–Shoenlein purpura (author’s translation). *Nouv Presse Med*. 1978;7:3913–4, 3919–20 (In French).