Recurrent and reversible acute renal failure in a patients with hematuria and Schönlein- Henoch purpura

E. Torregrosa de Juan, J. Hernández Jaras, J. J. Sánchez Canell, M. Pin, E. Tamarit, I. Rico, C. Calvo Gordo, H. García Pérez, R. Pons Prades, A. Rius and M. A. Fenollosa

Hospital General de Castellón.

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SUMMARY

Sumary Henoch- Schönelin purpura (HSP) is a small vessel vasculitis characterized by deposition of inmune complexes, mainly IgA and C3. It is a multisystem disorder affecting predominantly the skin, joints, gastrointestinal tract and kidneys. Clinical expression of nephritis ranges from transient isolated microscopic haematuria to rapidly progressive nephropathy. Acute renal failure is rare and is associated with episodes of macroscopic hematuria. These episodes are frequently associated with tubular damage and tubular obstruction by erythrocyte casts. We describe a patient with two episodes of acute renal failure after the onset of gross hematuria. Both episodes were reversible after six and four months respectively on hemodialysis.

Key words: *Henoch-Schönelin purpura. Acutre renal failure. Dialysis.*

RESUMEN

La purpura de Schönlein-Henoch es una vasculitis de pequeño vaso caracterizada por el depósito de inmunocomplejos, principalmente IgA y C3. Es un trastorno multisistémico que afecta predominantemente la piel, las articulaciones, el tracto gastro-intestinal y los riñones. A nivel renal la expresión clínica varía desde una microhematuria aislada transitoria, hasta el cuadro de nefropatía rápidamente progresiva. El fracaso renal agudo es raro y suele verse asociado a episodios de hematuria macroscópica. Estos episodios suelen cursar con daño y obstrucción tubular por cilindros eritrocitarios. En este caso clínico describimos un paciente que sufrió dos episodios de fracaso renal agudo reversibles precedidos por brotes de hematuria macroscópica y que precisaron hemodiálisis durante cuatro y seis meses respectivamente.

Palabras clave: Púrpura de Scholeïn-Henoch. Fracaso renal agudo. Hemodiálisis.

Correspondence: *Eduardo Torregrosa de Juan*
Hospital General de Castellón
Avda. Benicassin, s/n
12004 Castellón. España
torregrosa_edu@gva.es

INTRODUCTION

Henoch-Schönlein purpura is a small vessel systemic vasculitis characterized by deposition of immune complexes, mainly polymeric IgA and C3, in the skin, gastrointestinal tract, joints, and kidneys. Acute renal failure complicating Henoch-Schönlein purpura is usually due to a mesangial glomerulopathy with epithelial cell proliferation, crescent formation, and a greater or lesser degree of tubular damage. The clinical course of a patient with Henoch-Schönlein purpura and hepatic cirrhosis who suffered two episodes of acute renal failure separated in time that required replacement therapy for longer than six months and from which he recovered completely is reported. There are cases reported in the literature of acute renal failure secondary to episodes of gross hematuria in the setting of IgA nephropathy or Henoch-Schönlein purpura, but none of them had such severe and prolonged involvement, with two episodes at separate times.

CASE REPORT

A 42-year-old male patient attended the emergency room reporting erythematous, pruritic lesions in the lower limbs for the past two weeks, as well as gradual occurrence of edema, joint pain in malleoli and knees, and choluric urine. Personal history: The patient was a former intravenous drug user. He occasionally sniffed cocaine, drank heavily (2 liters of beer daily), and smoked two daily packs of cigarettes. Physical examination revealed pallor in skin and mucosal membranes, fetor hepaticus, vascular spiders in cheeks and upper hemithorax, bilateral parotid hypertrophy, two finger-breadth hepatomegaly, and purpuric lesions in lower limbs, some of them with central necrosis. Main laboratory data included: Hemoglobin 9.6 g/dL, ferritin 20 ng/mL, platelets 47,000, Quick index 66%, creatinine 2.6 mg/dL, urea 55 mg/dL, glucose 151 mg/dL, calcium 7.8 mg/dL, bicarbonate 20.9 mmol/L, total protein 6.5 g/dL, albumin 2.3 g/dL, proteinogram: polyclonal immunofixation, PTH 242 pg/mL, GOT 91 IU/L, GGT 55 U/L, GPT 73 U/L, IgG 2663 mg/dL, IgA 776 mg/dL. All other biochemical parameters were normal, including negative ANA, ANCA , and anti-BM, with normal C3 and C4 values. Urine analysis showed the presence of 1.4 g of protein

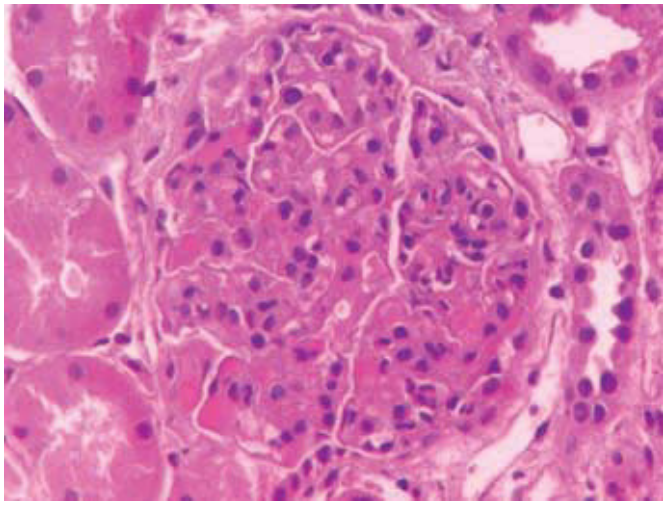

Figura 1.

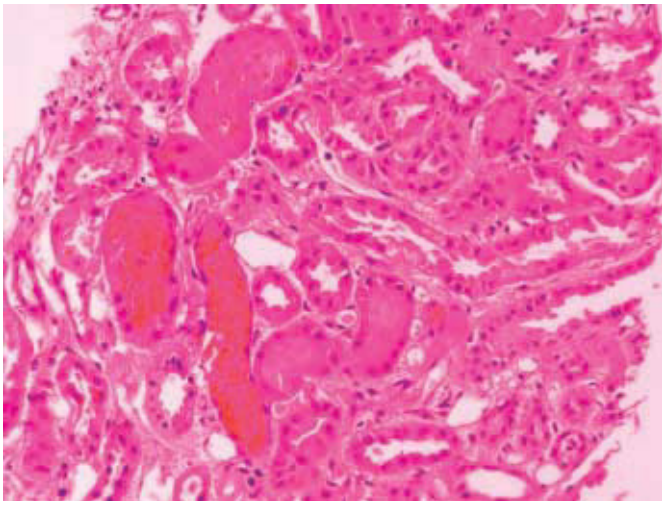

Figura 2.

and a sediment with abundant red blood cells per field. Hepatitis C serologic testing was positive, with a viral load of 156,000 IU/mL. As regards hepatitis B virus, there was positivity to anti-HBc IgG. HIV testing was negative. An abdominal ultrasound showed moderately enlarged kidneys with increased echogenicity. Hepatomegaly at the expense of the left and caudate lobe, with globally increased echogenicity. All other supplemental examinations (chest X-rays, echocardiogram, and ECG) were normal. After admission, patient was found renal function impairment (creatinine 3.3 mg/dL), intensification of gross hematuria, and arthromyalgia, mainly in upper limbs, with no skin lesions. Due to the impaired renal function and while waiting to be able to perform a renal biopsy, corticosteroid treatment was started as 3 daily boluses of 6-methylprednisolone 500 mg, followed by prednisone 60 mg/day for 8 additional days. This treatment improved the joint picture and stabilized kidney function (creatinine 3.5 mg/dL). Renal biopsy was performed on the sixth day of admission, and showed segmental endocapillary proliferation (IIb) in 4 of 10 glomeruli sampled, with a few polymorphonuclears without extracapillary proliferation, small vessel interstitial involvement, immunofixation positive for IgA, degenerative tubular changes with presence of intratubular blood casts, and focal tubular atrophy. All findings were consistent with a small vessel vasculitis such as Henoch-Schönlein nephritis. Based on the renal biopsy result and since no extracapillary proliferation was found, response to steroid treatment was poor and maintenance of such treatment involved risks because of liver disease, steroid treatment was discontinued after 8 days. At 29 days, the patient experienced a new purpuric outbreak in his lower limbs, with gross hematuria and severe kidney function impairment (creatinine 9.6 mg/dL), preceded by gastroenteritis induced by *Salmonella enteritidis*. Dialysis was started through a temporal catheter. During the rest of his hospital stay, the patient maintained urine output (1,000 mL/24 h) with persistence of gross hematuria. No renal function recovery was seen. Because of his clinical stability, he was discharged home to continue follow-up and ambulatory dialysis treatment. After hemodialysis for 7 months through a left radiocephalic fistula, the patient experienced a progressive renal function improvement, with remission of gross hematuria, and replacement therapy was therefore discontinued. During follow-up at the outpatient clinic, renal function recovery to creatinine values of 1.3 mg/dL, 24-hour proteinuria of 160 mg, and normal sediment was confirmed. The abovementioned hepatic changes persisted. Nineteen months after his initial admission, the patient again showed hematuria with no skin or joint clinical signs, associated to a systemic condition and severe acute renal insufficiency (creatinine 9.8 mg/dL, urea 215 mg/dL). Dialysis through an arteriovenous fistula was therefore restarted. Four months later, dialysis was again discontinued for renal function recovery (creatinine 1.9 mg/dL, urea 82 mg/dL). After one year of outpatient follow-up, the patient showed urea levels of 24 mg/dL and creatinine levels of 1.4 mg/dL, and urine analysis revealed microhematuria of 25-50 red blood cells/field, with no proteinuria.

DISCUSSION

Acute renal failure has often been associated to episodes of gross hematuria in patients with IgA nephropathy. However, this complication may also sporadically occur in patients with other nephritis such as Henoch-Schönlein purpura, postinfectious acute glomerulonephritis, focal necrotizing glomerulonephritis, and thin basement membrane disease.[1-4] In most cases, glomerular changes seen in the biopsy were not sufficient to explain renal function impairment. By contrast, the presence of red blood cells casts obstructing 40%-50% of tubular lumen and the marked tubular necrosis were the main findings made to explain the clinical picture.[1] Other mechanisms suggested to explain tubular necrosis and acute renal failure have included direct tubular toxicity induced by heme pigments or by intrarenal vasoconstriction resulting from binding of nitric oxide to molecules with a heme group.[5] In this regard, Sheerin et al showed that cells from the proximal convoluted tubule may phagocytize red blood cells, and that free iron and hemoglobin were toxic for these cells.[6] In our case, immunosuppressive treatment was started because of the presumptive diagnosis of rapidly progressive glomerulonephri-

tis. However, biopsy showed signs of tubular damage and presence of focal and segmental proliferation with moderate mesangial hypercellularity, without extracapillary proliferation, and immunosuppressive treatment was therefore discontinued, as in other cases reported in the literature.[7,8] However, the presence of multiple poor prognostic factors and severe renal insufficiency at diagnosis prompted the need for dialysis treatment for six months.[9,10] After this time, urine output gradually increased, and renal function recovered, which allowed for discontinuation of dialysis. This course is not uncommon, and has in fact been previously reported by other authors.[2] However, this case is unique in that it reports the course of a patient with Henoch-Schönlein purpura who experienced a second episode of acute renal failure caused by gross hematuria 19 months after the first episode from which he spontaneously recovered again, despite the existence of poor prognostic factors such as severe renal insufficiency at diagnosis, gross hematuria, proteinuria higher than 1 g, and interstitial infiltrate.

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