Case report

Henoch–Scho¨nlein purpura associated with human immunodeficiency virus infection

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Introduction

The occurrence of IgA nephropathy (IgAN) in patients with human immunodeficiency virus (HIV) infection is now well described [1]. It was first reported by Baunmelon et al. in 1989 [2], and the subject has recently been reviewed by Cohen [1].

In most reports the nephropathy has been mild with micro/macrosopic haematuria and minor proteinuria. However, there have been some patients with more severe renal disease [1]. There are three reports of Henoch-Scho¨nlein purpura (HSP) associated with HIV infection [3–5], of these none had all of the clinical criteria for HSP.

The renal biopsy changes reported in these HIV-infected patients are basically the same as those in patients with primary IgAN, with the additional finding on electron-microscopy of tubuloreticular structures in glomerular and epithelial cells [1].

We report a patient with classical HSP and HIV infection to further strengthen the link of HIV as a cause of secondary IgAN.

Case report

A 41-year-old male with severe haemophilia A (Factor VIII C level <1%) presented to hospital in October 1996 with acute abdominal pain. Examination revealed generalized peritonism but elicited no other significant findings. Unremarkable initial investigations included a full blood picture, urea and electrolytes, serum creatinine, liver function tests, calcium, amylase and plain abdominal radiography. Abdominal computed tomography (CT) scanning revealed free intraperitoneal fluid consistent with intraperitoneal bleeding.

He was treated with anti-haemophilia factor (AHF) and subcutaneous morphine.

His past medical history included: iatrogenically acquired human immunodeficiency virus (HIV) infection diagnosed in 1984 with p24 antigenaemia and low but stable T-helper CD4 lymphocyte counts without opportunistic disease; polymerase chain reaction positivity for the hepatitis C virus (HCV) with persistently normal hepatic transaminase parameters; multiple haemarthroses; post traumatic splenectomy.

G-CSF (Lenograstim) had been commenced in April 1995 to maintain his neutrophil count above 1.0 × 10^9/l whilst on zalcitabine. The dose of G-CSF on admission had been 263 μg weekly.

The symptoms slowly resolved and he was weaned off AHF. G-CSF was reintroduced because of persisting neutropenia. The following day a purpuric papular rash appeared on the lower limbs and palms. The patient reported that a similar rash had occurred frequently over the previous 9 months. He had noticed a temporal association to G-CSF and had assumed it a normal response to treatment. The rash always involved both ankles with arthralgia and in recent months had extended to the buttocks and palms. It invariably improved within 1–2 days but with residual scarring.

He further described episodic abdominal pain of similar character but increasing severity over 3 months prior to admission. G-CSF was discontinued.

Over the next month he experienced three further episodes of abdominal pain of increasing severity, recurrence of the rash (Figure 1), synovitis, and two episodes of macroscopic haematuria with dysmorphic red cells and casts. Records disclosed that microscopic haematuria and mild proteinuria had been present since 1986. He was not hypertensive. Serial CT scans revealed less intraperitoneal fluid but evolving small-bowel intramural oedema.

His haemoglobin concentration, serum urea, electrolytes, creatinine, liver function tests and C-reactive protein were consistently normal. Factor VIII levels were in the high therapeutic range. Antinuclear antibody screening was negative. The erythrocyte
sedimentation rate (ESR) peaked at 125 mm/h, serum complement components C3/4 decreased to 0.35/0.05 g/l (normal range 0.75–1.4/0.11–0.34) and serum immunoglobulin concentrations were IgA 23.0 g/l (0.64–3.4), IgM 3.95 g/l (0.3–1.7), IgG 52.2 g/l (5.8–13.7). Serum cryoglobulins were not detected. A skin biopsy (Figure 2) demonstrated leukocytoclastic vasculitis. Direct immunofluorescence revealed granular deposits of IgA and C3 consistent with Henoch–Schönlein purpura (Figure 3). A renal biopsy was not performed.

Discussion

The clinical diagnosis of HSP in this patient was based on the purpuric rash, arthralgia with synovitis, abdominal pain, and the presence of glomerular haematuria. Confirmation of the diagnosis in this case was by skin biopsy showing granular deposits of IgA and C3 and a leukocytoclastic vasculitis. Renal biopsy was not possible.

Immunopathological findings suggest that IgAN is the kidney-limited expression of a single disease spectrum with HSP as the systemic vasculitic form.

IgAN has been described in more than 20 patients with HIV [1] and the prevalence of covert disease in such patients is probably high. An autopsy evaluation of kidneys of AIDS patients has disclosed a 7.8% prevalence of mesangial IgA deposition [6].
The origin of mesangial IgA in primary IgAN is uncertain. One can usefully speculate as to the mechanisms responsible for the association of IgAN or HSP and HIV infection. Patients with HIV often have elevated serum IgA concentrations (as was the case in our patient), as do patients with alcoholic cirrhosis who also frequently develop secondary IgAN [7]. Analysis of circulating immune complexes from some patients with HIV and IgAN has shown IgA reactive with HIV p24 and gp41 [8]. Alternatively, the IgA in HIV patients has been described as polymeric IgA1 rheumatoid factor [9]. Studies of the immunochemistry of serum IgA in patients with primary IgAN suggest defective galactosylation of the hinge region of IgA1 that might be responsible for impaired clearance of IgA from the circulation and for its mesangial deposition [10]. The results of similar studies with HIV sera are awaited.

References


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