Case Report

Immune thrombocytopenic purpura presenting in an immunosuppressed patient after renal transplantation

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Case report

We report the case of a 30-year-old man with IgA nephropathy who had undergone a one-haplotype-matched renal transplant from his mother and who presented 9 months later with epistaxis and bruising after minimal trauma. He had been knocked several times whilst travelling to work in London and had noticed later that day that he had developed a large area of bruising over his left thigh. He had also had an episode of spontaneous epistaxis the previous day for the first time.

He had been well until 2 years previously when he had an episode of macroscopic haematuria and was noted to be significantly hypertensive with a blood pressure of 210/120 mmHg. Electrocardiography at the time showed left ventricular hypertrophy and examination of the urine demonstrated proteinuria and haematuria with dysmorphic red blood cells. Renal ultrasound examination revealed bilaterally small kidneys and a renal biopsy confirmed the diagnosis of IgA nephropathy.

The patient was subsequently non-compliant with antihypertensive therapy and his renal function continued to deteriorate until he was started on haemodialysis. He underwent a transplant from his mother 3 months later. Seven days post-transplant there was evidence on renal biopsy of mild cellular rejection which was treated with pulsed methylprednisolone (3 x 500 mg daily) and azathioprine was added to his regimen of prednisolone and cyclosporin. A further renal biopsy 1 month later showed no evidence of rejection, but he suffered from swinging pyrexias from weeks 4 to 7. The fever was typical of cytomegalovirus (CMV) infection, and there was mild transient elevation of hepatic enzymes, but cultures and serology for CMV and Epstein–Barr virus (EBV) remained persistently negative. Subsequently he was commenced on enalapril for hypertension along with amlopidine but had no other drug treatment in the intervening period except for a week long course of flucloxacillin. Otherwise he remained well with a urea of 6–7 mmol/l and a creatinine of 140–150 µmol/l until his admission with bleeding problems.

At this time on examination there was a large bruise on the left thigh and two smaller bruises in the right antecubital fossa from venesection. There were also scattered petechiae over his body but no retinal haemorrhage. Otherwise, examination was unremarkable. Routine blood tests showed a haemoglobin (Hb) of 12.3 g/dl, white cell count (WCC) of 6.8 x 10^9/l and a platelet count (plt) of 5 x 10^9/l. This compared with a platelet count of 262 x 10^9/l when last tested 16 days previously. A transfusion of 6 units of pooled platelets was given but failed to increase the platelet count (the post-transfusion level being 3 x 10^9/l).

Other causes of thrombocytopenia were excluded with a normal thoracic and abdominal CT scan and no serological evidence of underlying autoimmune disease. Bone marrow aspiration and trephine showed no evidence of infiltration but large numbers of megalakocytes including some juvenile forms, consistent with a diagnosis of ITP.

The patient was treated with prednisolone 60 mg daily instead of 12.5 mg daily and this resulted in a steady increase in the platelet count (Figure 1) until 1 month later it was stable at 180 x 10^9/l. The patient subsequently remained well and the dose of prednisolone was successively reduced after 1 month gradually to a maintenance of 7.5 mg daily with no relapse in the platelet count.

Discussion

ITP is a disease that normally presents as a primary entity in adults, although it may also be associated with an underlying immunoproliferative disorder or immunosuppressive disorder such as hypogammaglobulinaemia. It also occurs associated with human
Immune thrombocytopenic purpura after renal transplantation

Fig. 1. Platelet count (×10^9/L) vs time.

[Graph showing platelet count over time]

The underlying condition for which transplantation was performed in this case was idiopathic IgA Nephropathy (IgAN). This is itself a disease characterized by abnormalities in the systemic immune system. Histologically the renal lesions are characterized by the deposition of IgA within the mesangium of the glomerulus. There have been no consistent findings relating to the antigens involved although high IgA titres have been found to food proteins such as gliadin and casein, and bacterial and viral antigens. The systemic nature of the disease is demonstrated by the fact that it may recur 1–4 years after transplantation, and may be cleared if an IgA nephropathic kidney is inadvertently transplanted into a patient with renal failure for some other reason [8]. Some investigators have implicated autoimmune mechanisms involving the IgG system, observing high serum levels of IgG specific for the F\text{ab} region of autologous IgA in patients with idiopathic IgAN [9]. Autoantibodies specific for mesangial determinants have also been reported in some patients associated with increased levels during episodes of macroscopic haematuria [10]. These both suggest the possibility of autoantigenicity against glomerular antigens and F\text{ab} in IgAN.

In summary, we present a patient who developed ITP after a live-related donor renal transplant for IgAN despite the fact he was maintained on triple immunosuppression including prednisolone. It is not clear whether this is related to the process of transplantation or to autoimmune aspects of the patient’s underlying pathology; however, the immunogenic nature of the condition was convincingly demonstrated by the prompt response and remission induced by high-dose glucocorticoid therapy in addition to the maintenance treatment. We conclude, therefore, that in such cases high-dose corticosteroid treatment is appropriate and effective even if powerful immunosuppressive therapy is already employed.

References


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