

rence. Treatment for RVT has evolved from nephrectomy to thrombectomy and finally to thrombolytic therapy with anticoagulation, which is currently the standard treatment of choice [1].

Here, we present a young woman with bilateral RVT. Her case constituted the first clinical report of heterozygous MTHFR mutation with RVT, which was her only risk factor for the disease. MTHFR is a key enzyme for intracellular folate homeostasis and metabolism that catalyses the reduction of 5,10-methylenetetrahydrofolate to 5-methyltetrahydrofolate, the main circulating form of folate and the methyl donor for the vitamin B₁₂-dependent remethylation of Hcy to methionine. Results from previous studies showed that the 677TT MTHFR genotype can be considered as an independent risk factor for both arterial and venous thrombosis and may itself increase the risk for thrombosis even in the absence of other thrombophilic risk factors [3,4]. Recently, a novel MTHFR polymorphism, 1298A3C, which changes glutamic acid into an alanine residue, was shown to be associated with decreased enzyme activity but did not result in decreased folate plasma levels or increased plasma Hcy concentrations in homozygous or heterozygous members of neural tube defect families [5]. Also, others have reported that MTHFR 1298CC and MTHFR 1298AC had no effect on the risk for vein thrombosis [6]. In contrast, we present the first adult case that shows a relation between RVT and MTHFR-1298.

Conflict of interest statement. None declared.

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doi: 10.1093/ndtplus/sfq049

Advance Access publication 22 April 2010

A growing transplanted kidney

Sir,

Renal sinus lipomatosis (RSL) is a rare disease, described by replacement of parenchyma by sinus and/or perirenal fat. We report here an exceptional case of RSL on kidney graft after corticosteroid treatment was stopped, leading to functional graft transplantectomy.

A 74-year-old man received a first renal allograft in 2004. He was maintained on tacrolimus and mycophenolate mofetil with no rejection. Corticosteroid was stopped at the sixth month. The lowest serum creatinine level was 100 µmol/L. On February 2007, renal ultrasonography, performed for acute renal failure, showed dilatation of renal cavities. Contrast-enhanced computed tomography (CT) showed an enlarged kidney (14.3 cm) with an increased amount of fatty-like tissue in renal sinus and perirenal area (Figure 1). For 6 months, he experienced several obstructive renal failures and infections, leading to permanent nephrostomy. Biopsy of the sinus mass revealed an extensive fatty tissue with fibrosis. Finally, because of bilateral pulmonary embolism secondary to vena cava compression, persistent obstruction and continuous graft enlargement without conclusive histology, and despite good renal function, he underwent transplantectomy. Histological examination showed RSL (immunophenotypical labelling and biomolecular investigations negative for sarcoma) and atrophic chronic pyelonephritis graft.

RSL is essentially described on native kidneys [1,2]. Possible risk factors for RSL include ageing, obesity and pathologic states that cause renal inflammation such as intense corticotherapy and/or early rejection for transplants. Often, chronic or repeat urinary tract infections have been found, and one hypothesis is that RSL may be secondary to periodic leakages of urine into the peripelvic tissues [2]. RSL can also be part of replacement of destroyed or atrophic renal tissue [3]. It has also been suggested that high-dose steroid treatment by itself, Cushing's syndrome or obesity may have contributed to the development of fibrolipomatosis [4]. Intriguingly, a very few cases of RSL have been reported in the renal transplant population [5]. This case is intriguing for several reasons: The time between transplantation and the first symptoms was quite long (3 years). The recipient of the contralateral kidney did not develop any RSL. The patient experienced his first urinary tract infection at the same time as the first acute obstructive renal failure. No history of earlier acute rejection episodes has been found. He did not have a long-term steroid treatment and was not obese. Finally, our case is also



Fig. 1. CT shows extensive deposition of fat in the renal sinus of the transplant kidney (August 2007).

exceptional because it led to transplantectomy. Transplantectomy was decided in view of the rapidly increasing graft mass, responsible of vena cava compression and suspicion of malignity.

In conclusion, our case underlines that diagnosis of RSL, although exceedingly rare, should be considered when imaging exams demonstrate a fat-containing renal sinus mass. In transplant recipients, immunosuppressive regimen and malignancy risk underline the importance to establish differential diagnosis between fibrolipomatosis and fibrosarcoma, renal tumour or post-transplant lymphoproliferative disorder.

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Conflict of interest statement. None declared.

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doi: 10.1093/ndtplus/sfq061

Advance Access publication 28 April 2010

Doxycycline for haematopoietic stem cell transplantation-related thrombotic microangiopathy

Transplantation-associated thrombotic microangiopathy (TA-TMA) is a devastating consequence of allogeneic haematopoietic stem cell transplantation (HSCT) with a mortality rate of 60–90%. None of the interventions used, as used up till now in idiopathic thrombotic thrombocytopenic purpura (TTP) (fresh frozen plasma transfusion, plasma exchange and steroids), were effective to treat TA-TMA [1,2]. We report a dramatic improvement of TA-TMA in two HSCT patients [conditioning, cyclophosphamide, total body irradiation, graft-versus-host disease (GVHD) prophylaxis] using doxycycline.

A 36-year-old woman with Hodgkin's lymphoma received an allogeneic HSCT in December 2002. Twelve months later, she developed a biopsy-proven TMA (proteinuria, 3 g/day, microscopic haematuria, oliguric acute renal failure with creatinine level at 680 µmol/L; haemoglobin Hb, 6.3 g/dL; schistocytes; platelet count, $35 \times 10^9/L$; LDH, 1754 IU/L). The serum complement proteins were at normal levels, no mutations of the membrane cofactor protein were found and a plasma ADAMTS13 activity was found at 40%. Steroids, plasma exchange, fresh frozen plasma transfusion, vincristine and haemodialysis were tried with a partial response (haemoglobin, 7.3 g/dL, platelet $70\,000/mm^3$ both after treatment). Doxycycline 200 mg daily was added for a suspected gastrointestinal *Bartonella* infection. Within two months, haemoglobin and platelet count rose without transfusion to 10.8 g/dL and $234\,000/mm^3$, respectively. Despite improvement of haematological parameters, the patient remained dialysis-dependent. The second patient had a similar haematologic disease and course under doxycycline prescribed for a Bartholinitis.

Five patients with TTP and *Bartonella*-like erythrocyte inclusions, successfully treated with doxycycline, experienced recurrence of their TTP following cessation of treatment [3]. TA-TMA has a multi-factorial aetiology of endothelial damage. Doxycycline targeting the adherens junction on endothelial cells prevents vascular hyperpermeability [4]. Doxycycline as a potential treatment of TA-TMA warrants further studies.

Conflict of interest statement. None declared.

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doi: 10.1093/ndtplus/sfq072

Advance Access publication 26 April 2010

The spot urine protein/creatinine ratio is a simple, rapid and inexpensive method for monitoring patients with light-chain multiple myeloma

Protein electrophoresis of a 24-h urine collection (UPEP) is considered the standard method for following up patients with light-chain multiple myeloma [1]. The serum-free light-chain assay (SFLCA) has increasingly been used in this population [2], and in individual patients tracks well with proteinuria [3]. In addition, the SFLCA is also generally more sensitive than urine studies including immunofixation electrophoresis for detecting minimum residual light-chain disease [4,5]. However, the SFLCA is expensive, and due to inter-patient variation in the renal metabolism of light chains, the amount of proteinuria cannot be predicted by the SFLC concentration [1,5–7]. As proteinuria correlates better with renal dysfunction than SFLC and may be caused by factors other than light chains, serial measurement of urinary proteinuria is still considered essential [7].

The spot urine protein/creatinine ratio (SUPCR) has increasingly replaced the 24-h urine in patients with proteinuria from a variety of causes [8], but has not been examined in patients with multiple myeloma. As free light chains have a half-life of 2–6 h [9], the SUPCR is theoretically ideally suited to measure response to treatment within days of beginning therapy, and moreover, can be inexpensively and serially measured with rapidly available results. In this report, five patients with predominantly light-chain multiple myeloma were followed up by SUPCR and SFLCA. In Patient 1 and 2 (Figure 1A and B), progressive disease and subsequent response to therapy were accurately detected by SUPCR and in agreement with changes in the SFLCA. In Patient 3 (Figure 1C), bortezomib