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Focal segmental necrotising glomerulonephritis associated with selective IgA deficiency: two case reports

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Introduction: Selective IgA deficiency (SIgAD) is the most common primary immune deficiency worldwide. Clinical manifestations may vary, ranging from asymptomatic to recurrent infection, gastrointestinal disease, atopy and autoimmune disease. A variety of autoantibodies may develop, which do not necessarily correlate to clinically apparent disease. Of those that do, systemic lupus erythematosus and rheumatoid arthritis are most common, which often have renal sequelae. However, the occurrence of an isolated glomerulonephritis (GN) in the context of SIgAD, without the diagnosis of an autoimmune condition, is rare, and largely on a case by case basis. The most common pathologies seen are diffuse proliferative GN or mesangioproliferative GN, and less frequently minimal change disease or membranous nephropathy. There has been one previously published case report of an anti-neutrophil cytoplasmic antibody (ANCA) associated GN developing with SIgAD. Here, we describe two cases of a rapidly progressive, focal segmental necrotising GN developing with SIgAD, positive for not only ANCA but also numerous other autoantibodies.

Cases: Both patients presented with a rapidly progressive acute kidney injury and haemtoproteinuria on urinalysis, and were found to have SIgAD. Patient 1, a 69 year old female, presented with a creatinine of 348 umol/L and was predominantly positive for myeloperoxidase (MPO) on ELISA, in addition to various other autoantibodies (table 1). Renal biopsy confirmed a focal segmental necrotising GN, and she was started on methylprednisolone and cyclophosphamide. She was continued on oral azathioprine and a reducing dose of prednisolone, and has made a good renal recovery with a current creatinine of 109 umol/L. Patient 2, a 79 year old male with a history of pulmonary fibrosis, presented with a more insidious 12 month history followed by a sudden acute kidney injury, with creatinine rising from a baseline of 160 umol/L to 1116 umol/L. He required initiation of haemodialysis, and renal biopsy again confirmed a focal segmental necrotising GN with extraglomerular vasculitis. He was previously found to have various raised autoantibodies whilst undergoing investigation for pulmonary fibrosis, and these remained largely unchanged (table 1). He was pulsed with methylprednisolone, followed by a weaning course of oral prednisolone, and treatment with cyclophosphamide is ongoing. He remains on haemodialysis, however has an increasing urine output and renal recovery is expected.

Discussion: The combination of SIgAD and intrinsic renal disease not due to an underlying autoimmune disease is rare, and although other pathologies have been reported, there is only one previous publication of a necrotising GN with several positive auto-antibodies, including ANCA. The treatment of SIgAD is usually focused on managing the complications, for example antibiotic therapy or management of autoimmune disease. However, the increasing number of case reports describing GN associated with SIgAD raises the question of whether monitoring of these patients, such as regular urinalysis, should occur. The purpose of our report is therefore to raise awareness of the association of SIgAD with GN, describe the very rare occurrence of a focal segmental necrotising GN with several positive auto-antibodies and SIgAD, and discuss potential management strategies for the future.