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A silent storm: report of a case of progressive renal failure secondary to IgG4-related kidney disease

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IgG4-related disease is an immune-mediated fibroinflammatory condition that can affect multiple organs. With multi-system symptoms, it can disguise itself as several diseases. Recent years have seen an increase in awareness of this condition. However, there is some way to go to improve early recognition and treatment of this entity.

We report the case of a 74 year old male ex-quarry worker with declining renal function, difficult to treat hypertension and proteinuria. He reported fatigue, joint pains, and a transient rash on his legs 1 month before presentation. He was euvolaemic on examination. Multiple hyperpigmented papules were present on his back. There was a trace of blood in his urine. Initial tests revealed mild proteinuria, hypocomplementaemia, a high ANA titre, an elevated Rheumatoid factor and a high plasma viscosity. A recent ultrasound showed normal looking kidneys. His eGFR had been slowly declining from normal since the beginning of 2015 and then took a nosedive from the beginning of 2019. It was 24ml/min/1.73m2 in clinic. A renal biopsy was performed 3 days later.

His past medical history revealed a diagnosis of silicosis which spawned from a protracted work-up for progressive dyspnoea in early 2015. A biopsy commented on a degree of silicosis. However, this did not account for all the features. Special stains for mycobacteria and fungi were performed, but did not identify any such organisms. In July 2016, he presented with retroorbital pain, diplopia, and what seemed like 4th and 6th nerve palsies on examination, eventually culminating in a diagnosis of Tolosa-Hunt Syndrome. An MRI of his head and orbit was normal.

MPO, PR3, and anti-GBM antibodies were absent. Antibodies to dsDNA were absent. Alpha-galactosidase levels were normal. His kidney biopsy showed a plasma cell rich tubulo-interstitial nephritis and accompanying storiform interstitial fibrosis. This was highly suggestive of an IgG4-related tubulo-interstitial nephritis. There was no significant staining on immunofluorescence. Although there was a modest elevation of his serum IgG, IgG4 subclass levels were normal. Prednisolone was started.

We are unsure how much, if any, of his past medical history can be linked to his renal pathology and if it can all be unified with a diagnosis of IgG4-related disease. The histological appearances are highly suspicious for IgG4-related renal disease. On recent follow-up, he demonstrated a favourable response to steroids with an eGFR of 34ml/min/1.73m2. However, it must be borne in mind that his renal function had declined significantly by the time he was assessed, and that a modest improvement may be all that can be hoped for.

IgG4-related disease can present with subtle symptoms and affect multiple organs, making it difficult to diagnose. There was no history or evidence of autoimmune pancreatitis, sclerosing cholangitis, or salivary gland involvement in our patient, which are common associations of IgG4-related disease. It is imperative that clinicians consider this differential in patients such as the one described here. Early treatment with steroids results in a characteristic improvement, and unless this window of opportunity is taken advantage of, outcomes can be poor.