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Evidence for IgG4 and ANCA disease association. A case study of MPO positive vasculitis with IgG4 positive plasma cells on renal biopsy in a patient with a background of pancreatic IgG4 disease.

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Background
Both IgG4 related disease and ANCA vasculitis can affect several different organs and both may present with general systemic symptoms such as fever, weighloss mylaigia but also lung, renal and GI involvement. Both are rare conditions.

Case Presentation
We present a case report of a 64 year old woman. She has a past medical history of Graves diseae, Cerebrovascular disease and Diabetes. She presented in 2017 to the hepatobiliary team with loose stool and abdominal pain. Endoscopic ultrasound showed a dilated CBD proximal to a stricture and an abnormal pancreas. A biopsy showed a chronic pancreatitis with probable IgG related autoimmune pancreatitis. Her IgG4 titres were raised but as she remained stable she was not treated with corticosteroids. Her creatinine at this time was 61mmol/l with an eGFR >90.

In September 2019 she presented to the Gastroenterology outpatient clinic feeling unwell. She was admitted to a local hospital with raised inflammatory markers and an acute kidney injury (AKI) with a creatinine of 200mmol/l. She was transferred to the renal unit for further investigation. She had symptoms of sinus pain, myalgia, shortness of breath and a cough. Her IgG4 levels were raised at 5.73 g/l (normal < 1.35 g/l). However her MPO level was now positive at >8 with a PR3 <0.2. Her urine dip showed 1+ protein and 2+ blood.

She proceeded to a renal biopsy which demonstrated features consistent with a pauci-immune crescentic glomerulonephritis with segmental necrotising lesions and approximately 65% crescents. In addition, there were significant numbers of IgG4 positive plasma cells, with an IgG4/IgG ratio of greater than 40%, in keeping with renal involvement by the known IgG4 related disease. There was moderate chronic tubulointerstitial damage.

She received IV methylprednisolone followed by oral Prednisolone 60mg daily prior to the biopsy along with Cyclophosphamide.

During her admission she developed a left hemiparesis and expressive dysphasia. Her MRI head showed bilateral acute infarcts in multiple territories with established old infarcts. MRA showed no intracranial vessel abnormality.

Outcome
Gradually her neurological symptoms and renal function continued to improve. Her latest results show an improved creatinine of 170mmol/l with eGFR 27. Her MPO titre has reduced to 5.0 and IgG4 is now within normal range at 0.98.

Discussion

This is a case of combined clinical and histological features of an ANCA associated crescentic glomerulonephritis and IgG4 related disease. There is only a few published reports of similar IgG4 and ANCA positivity however this appears to be the first case with both histological changes being present in the kidney.