Reply

Thank you for your comments in relation to our recently published article "A Case of IgG4- Related Retroperitoneal Fibrosis (IgG4-RPF) with Significant Involvement of the Abdominal Aorta — a Clinical and Diagnostic Challenge."[1]

In relation to the first comment regarding the lack of data for the patient's dark urine discoloration, the patient presented complaining of flank pain and a change in urine color, concerned that there was blood in his urine. A urinalysis was performed that demonstrated the presence of red cells, confirming hematuria. We observed a rise in the patients creatinine in keeping with an acute kidney injury. A computed tomography angiogram demonstrated extensive para-aortic soft tissue changes causing ureteric compression and secondary hydronephrosis owing to left-sided ureteric obstruction. Subsequently, a ureteric stent was inserted and the hematuria cleared up.

We did not suspect intrinsic renal IgG4-related disease, but we are aware that renal involvement in IgG4-related disease occurs in 15% of patients.[2] We agree that gross hematuria is a rare clinical finding in IgG4-related disease. However, IgG4-related diseases can affect multiple organs, including the kidneys; IgG4-related kidney disease or tubulointerstitial nephritis secondary to IgG4 disease and can present with hematuria, proteinuria, hypocomplementemia, and chronic renal failure.[2] In the context of IgG4-related disease, the kidney may be affected directly by histopathological lesions affecting the parenchyma or indirectly by retroperitoneal fibrosis (RPF), which causes obstruction of the urinary tract and renal impairment. There are reports of patients with IgG4-RPF presenting with flank pain, obstructive urinary symptoms, and peripheral oedema owing to ureter encasement by fibrosis, similar to our patient.[3-5]

We agree with the second comment that statin-induced pancreatitis is a rare form of drug-induced pancreatitis. When our patient had acute pancreatitis, the assessment was performed by a different medical team, which concluded that this episode was related to simvastatin use. Clinical documentation states other causes such as gallstones, alcohol use, hypercalcemia, and pancreatic malignancy were excluded. We note that IgG4 levels had not been evaluated. Retrospectively, we suspect this previous episode of pancreatitis 18 months prior was indeed IgG4 related; autoimmune pancreatitis is a common presenting condition of IgG4-related disease.

The third comment refers to a patient that the authors treated three times before the diagnosis of IgG4-RPF was made. We note this patient was initially treated with steroids only, achieved remission, but surveillance magnetic resonance imaging subsequently suggested active features of RPF and mycophenolate mofetil (MMF) was added to the treatment regime. In relation to our patient, we had a strong suspicion of IgG4-RPF early during their admission, which resulted in targeted investigations being performed. Histology confirmed our suspicion of IgG4-RPF in a short period of time and our patient was commenced on MMF at the same time as steroids. This strategy allowed for early weaning of steroids to minimize their long-term side effects and decrease the likelihood of relapse with steroid tapering. Relapse is common in IgG4-RD, particularly with the reduction of glucocorticoids and the 2-year relapse rates are 23.0% to 41.4%.[6] Our patient is currently on a tapering dose of steroids and MMF. He had a recent follow-up and surveillance imaging at 24 months, which suggests no reoccurrence of IgG4-RPF features clinically or radiologically. We note that, although the pathological diagnostic criteria for IgG4-RPF have been established, the pathological features associated with disease relapse are not well-reported.[6]

Sinead Gormley, MBBChBAO

Department of Vascular & Endovascular Surgery
Waikato Hospital
Waikato, New Zealand

Faculty of Medical & Health Sciences
University of Auckland
Auckland, New Zealand

Paola Tacuri Bravo, MBBChBAO

Department of Histology
Waikato Hospital
Waikato, New Zealand

Xavier Kos, MBChB, FRACR

Department of Interventional Radiology
Waikato Hospital
Waikato, New Zealand

Kamal Solanki, MBBS, FRACP

Department of Rheumatology
Waikato Hospital
Waikato, New Zealand

Manar Khashram, MBChB, PhD, FRACS

Department of Vascular & Endovascular Surgery
Waikato Hospital
Waikato, New Zealand

Department of Surgery
University of Auckland
Auckland, New Zealand

REFERENCES

1. Gormley S, Bravo PT, Kos X, Solanki K, Khashram M. A case of IgG4-related retroperitoneal fibrosis with significant involvement of the abdominal aort—a clinical and diagnostic challenge. J Vasc Surg Cases Innov Techn 2022;8:275-80.

2. Salvadori M, Tsalouchos A. Immunoglobulin G4-related kidney diseases: an updated review. World J Nephrol 2018;7:29-40.
3. Wallwork R, Wallace Z, Perugino C, Sharma A, Stone JH. Rituximab for idiopathic and IgG4-related retroperitoneal fibrosis. Medicine 2018;97:e12631.
4. Perugino CA, Wallace ZS, Meyersohn N, Oliveira G, Stone JR, Stone JH. Large vessel involvement by IgG4-related disease. Medicine 2016;95:e3344.
5. Wang K, Wang Z, Zeng Q, Zhu L, Gao J, Wang Z, et al. Clinical characteristics of IgG4-related retroperitoneal fibrosis versus idiopathic retroperitoneal fibrosis. PLoS One 2021;16:e0245601.
6. Zongfei J, Lingli C, Ying S, Lingying M, Lijuan Z, Dongmei L, et al. Clinical and pathological predictors of relapse in IgG4-related disease. Arthritis Res Ther 2022;24:106.

https://doi.org/10.1016/j.jvscit.2022.07.006