Immunoglobulin G4-related disease with large-size vessel involvement is more diverse than originally thought

We read with great interest the article by Gormley et al,[1] which focused on the diagnosis and management of a patient with immunoglobulin G4-related disease (IgG4-RD) affecting both the abdominal aorta and the common iliac arteries.[1] Although the findings of their reported case were thoroughly discussed, we believe that several issues should be highlighted and further elucidated.

First, although the authors had described in detail the clinical and laboratory findings, they provided no data regarding the dark urine discoloration. Gross hematuria, the most common cause of dark urine, is rarely seen in patients with retroperitoneal fibrosis and will usually be attributed to increased pressure in the pelvicalyceal system due to hydronephrosis.[2] However, in some cases of localized retroperitoneal fibrosis, gross hematuria resulting from compression and renal vein hypertension has been described.[3]

Second, the authors reported that the previous episode of acute pancreatitis (AP) had correlated with statin use. Statin-induced pancreatitis is a rare form of drug-induced pancreatitis and considered to be a diagnosis of exclusion.[4] However, the authors noted the slowly evolving nature of IgG4-RD and the long latency period preceding the definitive diagnosis.

In support of their findings, we have treated a patient with IgG4-RD who had developed periaortitis of the abdominal aorta and periarteritis of both renal arteries 2 years after the initial diagnosis. Similar to their patient, our patient had also had a history of three episodes of AP of unknown etiology before the diagnosis of IgG4-RD, which was established during a new episode of AP accompanied by renal insufficiency due to IgG4-related chronic tubulointerstitial fibrosis. We treated our patient with corticosteroids, and complete remission was achieved. However, the last magnetic resonance imaging study had revealed vessel wall thickening of the abdominal aorta and both renal arteries, compatible with IgG4-related periaortitis of the abdominal aorta and periarteritis (Fig). Hence, alternative therapeutic options were considered, and our patient received combined therapy with corticosteroids and mycophenolate mofetil.

In conclusion, IgG4-RD is a chronic, fibroinflammatory condition with multisystemic involvement occurring synchronously or metachronously. IgG4-RD seems to evolve insidiously and to result in a greater variety of patterns of organ involvement than previously thought, even when patients are deceptively considered to be in complete remission.

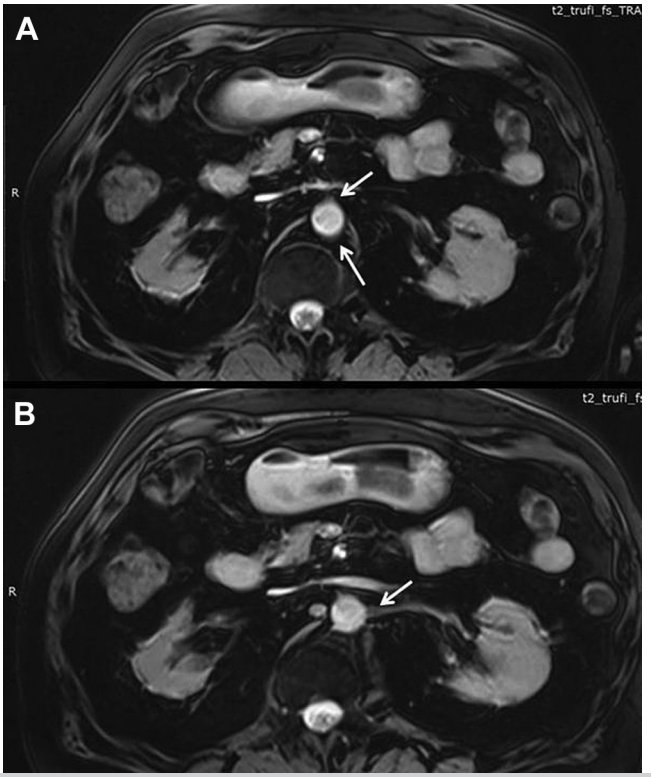

Fig 1. Periaortitis (**a**; *arrows*) and periarteritis of renal arteries (**b**; *arrow*) occurring metachronously in a patient with immunoglobulin G4-related disease (IgG4-RD).

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https://doi.org/10.1016/j.jvscit.2022.07.005