

**Letter to  
the Editor**

## Immunoglobulin G4-Related Aortic Disease

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and Satoshi Uesugi, MD**Keywords:** immunoglobulin G4, IgG4-related aortopathy, aortic dissection, aortic aneurysm

Uchida et al. published an article in 24 (2018) issue of the journal entitled “Immunoglobulin G subclass 4-related lymphoplasmic thoracic aortitis in a patient with acute type A aortic dissection,” in which they presented the case of a patient who was referred for surgical treatment.<sup>1)</sup> Histopathologic examination of his aortic tissue confirmed the diagnosis, showing marked adventitial thickening with fibrosis and an IgG4-positive plasma cell infiltrate.

Early in 2018, we also reported the case of a 65-year-old male patient with an enlarged ascending aorta and type B aortic dissection.<sup>2)</sup> The patient had experienced retroperitoneal fibrosis, membranous nephropathy, and acute coronary syndrome. Serum IgG4 levels were not measured and a pathological examination did not indicate that the retroperitoneal fibrosis and membranous nephropathy were IgG4 related. A 2-year course of steroid therapy was prescribed and contributed to recovery from the retroperitoneal fibrosis and nephropathy. The patient’s IgG4 level after the therapy was 19.8 mg/dL. He did well for several years without steroid therapy but then developed type B aortic dissection 3 years ago, at which point his serum IgG4 level had risen to 190 mg/dL. Chest computed tomography showed the dissection associated with thickened periaortic changes.

Despite its importance for postoperative disease management, a definitive diagnosis of IgG4-related disease is not easy to obtain preoperatively.<sup>3)</sup> IgG4-related inflammation can induce morphological changes in the aorta. For example, Lindsay et al.<sup>4)</sup> presented a case of periaortitis with periaortic soft tissue surrounding the abdominal aorta. We wondered whether Fig. 1 of Uchida et al. actually shows periaortic thickness suggesting IgG4-related disease. It would therefore be informative to know the perioperative serum IgG4 levels of their patient for IgG4-related diseases, steroid therapy is often prescribed for IgG4-related diseases, but the required therapeutic dose differs on a case-by-case basis. Tajima et al.<sup>5)</sup> have suggested that higher doses of corticosteroids might be required for the treatment of IgG4-related cardiovascular diseases compared with other IgG4-related diseases. We are prompted to ask: did Uchida et al. consider steroid therapy for their patient?

In conclusion, IgG4-related disease is a new clinical entity that can affect a variety of organs. IgG4-related aortic diseases cause various morphological changes; however, clinical features and pathological findings can aid in obtaining definitive diagnoses when different types of aortopathy are implicated.

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### Disclosure Statement

None declared.

### References

- 1) Uchida T, Hamasaki A, Kuroda Y, et al. Immunoglobulin G subclass 4-related lymphoplasmacytic thoracic aortitis in a patient with acute type A aortic dissection. *Ann Thorac Cardiovasc Surg* 2017; doi: 10.5761/atcs.cr.17-00111.

- 2) Sugaya A, Misawa Y, Ohki S, et al. A case of immunoglobulin G4-related aortic diseases. *Clinical Case Reports* 2018; **6**: 606-8.
- 3) Misawa Y. Immunoglobulin G4-related cardiovascular diseases. *Ann Thorac Cardiovasc Surg* 2017; **23**: 281-5.
- 4) Lindsay D, Ismajli M, Bucknall R, et al. Simultaneous presentation of IgG4-related chronic peri-aortitis and coeliac disease in a patient with Marfan's syndrome. *Rheumatology (Oxford)* 2016; **55**: 1141-3.
- 5) Tajima M, Nagai R, Hiroi Y. IgG4-related cardiovascular disorders. *Int Heart J* 2014; **55**: 287-95.