Early Detection and Intervention of Coronary Artery Involvement in Immunoglobulin G4-related Disease

Key words: IgG4-related disease, coronary artery

We wish to express our gratitude for your insightful comments regarding our case report (1). We agree with your comment that IgG4-related coronary periarteritis has been either overlooked or underdiagnosed in many patients with inflammatory abdominal aortic aneurysm (AAA). We therefore emphasized the utility of echocardiography in assessing coronary periarteritis in patients with IgG4-related diseases, even when they did not present with any cardiovascular symptoms.

If we assume there are some asymptomatic patients who have IgG4-related diseases without ischemic conditions, we cannot conclude that such patients would experience better outcomes at the time of the initial diagnosis because the clinical course of IgG4-related diseases can vary widely. We must at least control the disease activity caused by inflammation.

In general, corticosteroid therapy is effective for arteritis (2), particularly among IgG4-related diseases. However, we recognize that the efficacy of corticosteroids in patients with IgG4-related arteritis periarteritis is controversial. Ruggio et al. reported a patient with IgG4-related coronary periarteritis causing myocardial infarction; the subsequent induction of corticosteroid, antiplatelet, and oral anticoagulation therapy resulted in the patient’s long-term stability (3). In contrast, Nishimura et al. reported a patient with IgG4-related coronary periarteritis; despite balloon angioplasty performed successfully without induction of steroid therapy, the aneurysms and focal stenosis progressed after the intervention. However, steroid treatment considerably improved the blood flow and did not exacerbate the aneurysms (4). These cases indicate that corticosteroids are essential and effective in patients with IgG4-related coronary arteritis. However, as we mentioned in our review (1), there were some cases in which corticosteroids were ineffective; in those cases, stenosis and/or aneurysms were present before treatment (1). Kanzaki et al. reported a patient with IgG4-related coronary periarteritis who underwent coronary bypass surgery without corticosteroid therapy. Five years after the coronary bypass surgery, ¹⁸F-fluorodeoxyglucose (FDG) positron emission tomography revealed an elevation in the FDG uptake in coronary periarteritis (5). The persistent inflammation of the coronary artery could not be improved by surgical intervention (5). These cases show that surgical intervention or corticosteroids solely might be not enough for patients with stenosis or aneurysms and indicate that corticosteroids are necessary to improve inflammation in patients with IgG4-related coronary periarteritis, with surgical interventions, such as bypass grafting, occasionally needed. In summary, we believe that corticosteroid therapy is essential for controlling the disease activity of patients with IgG4-related periarteritis. If corticosteroids are ineffective, surgical interventions are necessary in patients with IgG4-related periarteritis (5). Our opinion is based on our experience and previous case reports (1, 3-5). We feel that increasing the number of subjects would help clarify this issue. Further investigations are thus needed in order to establish an optimal therapeutic strategy in the future.

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