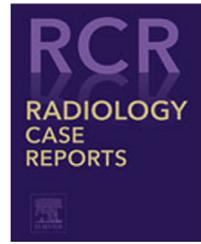


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Case report

Therapeutic response of immunoglobulin 4-related aortitis and pancreatitis demonstrated by diffusion-weighted MRI

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ABSTRACT

Immunoglobulin (IgG) 4-related disease is a systemic inflammatory disease, and it affects vascular system as aortitis, periaortitis, or aneurysm. However, due to a lack of serum biomarker on aortic damage and the multiorgan involvement, it is difficult to assess aortic inflammatory activity of IgG4-related disease. We described a case of IgG4-related pancreatitis and aortitis, which was visualized with magnetic resonance merged image of diffusion weighted and T₁ weighted images. The aortic signal intensity or apparent diffusion coefficient value reduced or increased after oral prednisone administration, respectively. Magnetic resonance diffusion weighted image and apparent diffusion coefficient may be a useful imaging tool for assessment of vascular inflammation in IgG4-related aortitis.

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Introduction

Immunoglobulin (IgG) 4-related disease is a systemic inflammatory disease characterized by increases of serum IgG4, a dense lymphoplasmacytic infiltrate rich in IgG4 plasma cells and fibrosclerosis. It may affect vascular system as aortitis,

periaortitis, and aortic aneurysm [1]. Yabusaki et al [2] visualized vascular inflammation in IgG4-related aortitis/periaortitis with ¹⁸F-fluorodeoxyglucose positron emission tomography/computed tomography (¹⁸F-FDG-PET/CT). However, high cost, radiation exposure, and availability are serious limitations of ¹⁸F-FDG-PET/CT. Magnetic resonance diffusion weighted imaging (DWI) can visualize diffusion capacity of

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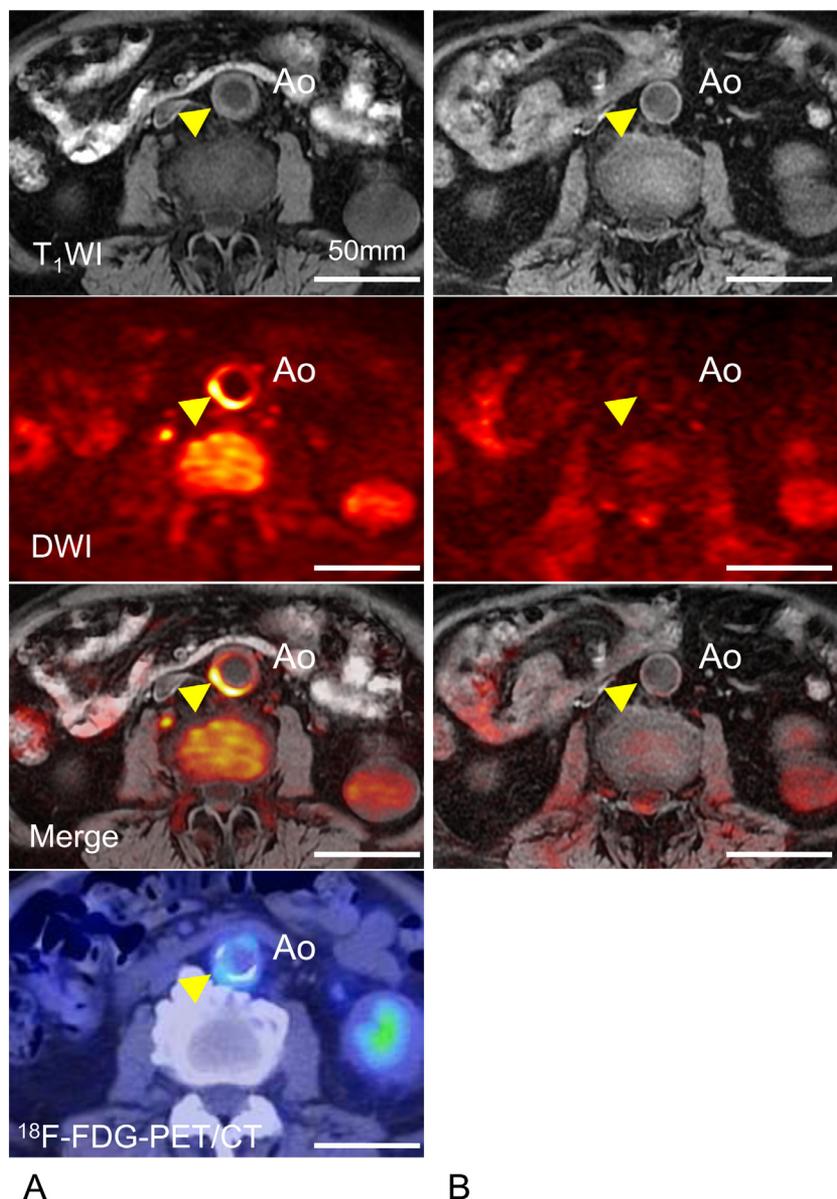


Fig. 1 – Response to 8-week course of systemic steroid therapy in 80-year-old man with IgG4-related aortitis and pancreatitis. (A) Axial T₁WI with fat suppression before treatment shows marked thickening of the abdominal aorta (arrow). Diffusion weighted imaging (DWI) and merged images of DWI and T₁WI show high signal intensity in the right to posterior portion of the aortic wall (arrow). ¹⁸F-FDG-PET/CT shows increase in FDG uptake in the right to posterior wall (arrow). (B) The aortic thickening improved 8 weeks after oral prednisone administration (30 mg/day) (arrow). The aortic high signal intensity on DWI disappeared after the treatment (arrow).

water molecules, and was used to detect the active phase of Takayasu arteritis [3]. Therefore, we applied this technique to visualize aortic inflammatory activity in a patient with IgG4-related disease.

Case report

A 80 years-old male was referred to our hospital for a detailed examination on the elevation of pancreatic enzymes.

The laboratory data showed an elevation of pancreatic amylase (201 IU/L), lipase (145 IU/L), IgG (3031 mg/dL) and IgG4 (1800 mg/dL). Ultrasonography showed swelling of pancreas and abnormal thickening of abdominal aorta. To evaluate the abnormalities, we performed noncontrast T₁ weighted image (T₁WI), and DWI (b values, 0 and 1000 mm²/s) (Signa HDxt 1.5T, GE Healthcare) and ¹⁸F-FDG-PET/CT. Axial T₁WI with fat suppression showed marked thickening of the abdominal aorta (Fig. 1A). Merged images of DWI and T₁WI showed high signal intensity on DWI in the pancreas (Fig. 2A) and aortic wall (Fig. 1A). ¹⁸F-FDG PET/CT also showed increase in FDG uptake

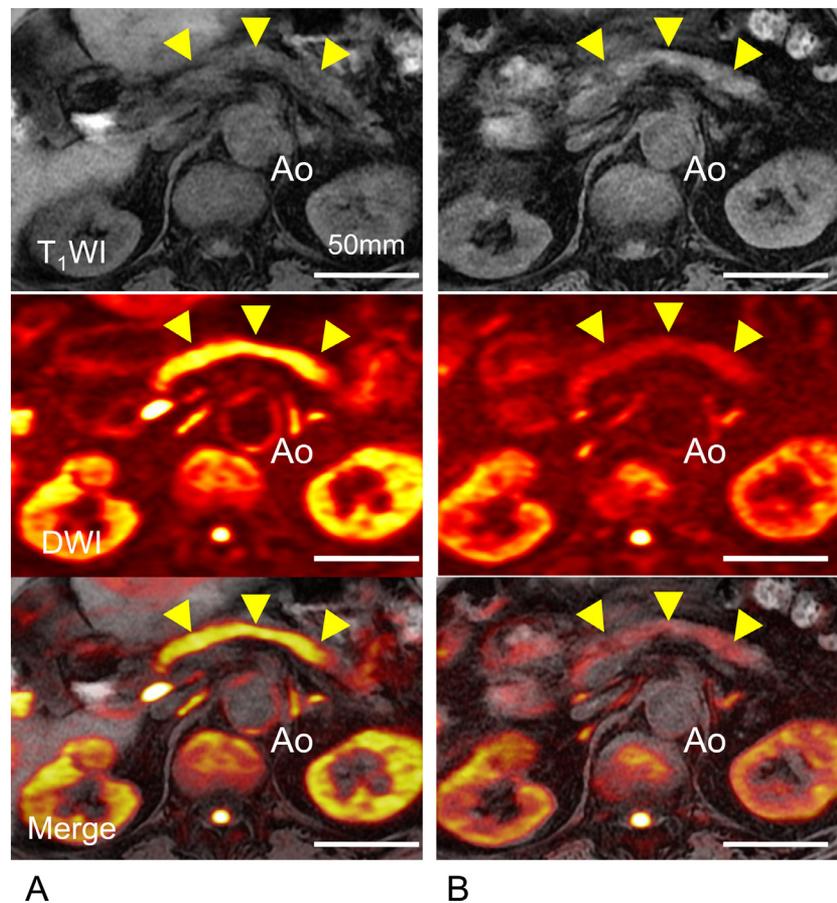


Fig. 2 – Response to 8-week course of systemic steroid therapy in 80-year-old man with IgG4-related aortitis and pancreatitis. (A) Axial Diffusion weighted image (DWI) and merged images of DWI and T₁WI before treatment show high signal intensity in the pancreas (arrows) and abdominal aorta. Kidney, spinal cord, and vertebral bone marrow show physiological high signal intensity on DWI. (B) The pancreatic high signal intensity on DWI disappeared after oral prednisone administration (30 mg/day).

in the aortic wall (Fig. 1A). Based on the findings, the patient was diagnosed as IgG4-related pancreatitis and aortitis. Oral prednisone administration (30 mg/day) for 8 weeks reduced the serum levels of IgG4 (172 mg/dL). T₁WI after the medication showed reduction of the aortic thickening (Fig. 1B) and disappearance of the pancreatic (Fig. 2B) and aortic (Fig. 1B) high signal intensity on DWI. The apparent diffusion coefficient (ADC) value of the aortic wall after the treatment ($2.0 \pm 0.4 \times 10^{-3} \text{ mm}^2/\text{s}$) was larger than that before the treatment ($1.5 \pm 0.3 \times 10^{-3} \text{ mm}^2/\text{s}$, $n = 22$, $P < .0001$).

Discussion

IgG4-related disease is a systemic inflammatory sclerosing disease, and a proportion of the patients may have aortic lesions. Yabusaki et al [2] evaluated 37 patients with IgG4-related disease with ¹⁸F-FDG-PET/CT. Fifteen (41%) patients exhibited IgG4-related aortitis, and (80%) of them showed multiple region involvement. The most common site was

the iliac arteries (35%), followed by the infra-renal abdominal aorta (33%), and thoracic aorta (8%). Perugino et al [4] examined 160 patients with histologically proven IgG4-related disease. Thirty five (22%) had large vessel involvement as primary IgG4-related vasculitis and aortitis and/or periaortitis secondary to retroperitoneal fibrosis. The most common site of the primary vasculitis was the thoracic aorta (62%), followed by the abdominal aorta (31%), and carotid artery (23%). The present case has synchronous lesions in pancreas and abdominal aorta. Based on the clinical and MRI findings, it is considered primary IgG4-related aortitis rather than periaortitis secondary to retroperitoneal fibrosis.

We demonstrated aortic inflammation and its resolution with DWI in patients with IgG4-related aortitis. As diffusion imaging is limited in spatial resolution, we merged the diffusion images with the T₁WI to better localize the high signal intensity in the thickened aorta and pancreas. This is a first report on noninvasive assessment of aortic inflammation in IgG4-related aortitis without radiation exposure and contrast agents. Kamper et al [5] examined 16 patients with chronic periaortitis with MRI and ¹⁸F-FDG-PET/CT, and found

a strong negative correlation between ADC and standardized uptake value of ^{18}F -FDG. The chronic periaortitis indicates a group of idiopathic fibro-inflammatory diseases, usually located around the infrarenal portion of the abdominal aorta and the iliac arteries [6]. It is considered that IgG4-related disease accounted for 30%-60% of idiopathic retroperitoneal fibrosis [6,7]. Although the authors did not mention the IgG4-related disease in the chronic periaortitis study [5], the cohort could include IgG4-related periaortitis secondary to retroperitoneal fibrosis. The present case and the previous study suggest a usefulness of DWI in IgG4-related aortitis/periaortitis.

IgG4-related aortitis is histologically characterized by adventitial lymphoplasmacytic infiltration, lymphoid follicle formation, obliterative adventitial phlebitis, and fibrosis [8]. Therefore, increased ^{18}F -FDG uptake in the aortic wall may reflect an abundance of chronic inflammatory cells in the lesion. However, the nature of the impaired diffusion capacity in the aortic wall on DWI is unclear. It is necessary to be further investigation.

We assessed regional inflammatory activity in IgG4-related aortitis with ADC values. The arterial ADC values after treatment were larger than before the treatment, comparable with findings of a previous study of chronic periaortitis [5]. Because the ADC values may be affected by acquisition sequences, the value can use a marker for follow-up assessment of lesions in each patient who are imaged on the same scanner with identical sequences.

Because magnetic resonance DWI does not require radiation exposure and contrast agents, it may be a useful imaging tool for assessment of vascular inflammation in IgG4-related aortitis.

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