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Case Report

A Case of Ruptured Immunoglobulin G4-Related Periaortitis

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An 80-year-old man had high serum immunoglobulin G4 (IgG4) concentration and fibrous thickening of the soft tissue mass surrounding the region from the abdominal aorta to the bilateral iliac arteries, suggestive of IgG4-related periaortitis. He presented to our emergency department with sudden-onset abdominal pain and lumbago. Computed tomography revealed a ruptured abdominal aorta. He was a poor candidate for open surgery due to his hostile abdomen. Therefore, endovascular aneurysm repair was performed. No consensus about the surgical indication for IgG4-related arterial disease has been reached yet. We believe that a novel indicator is needed for this disease.

Keywords: inflammatory abdominal aortic aneurysm, aortic disease, endovascular surgery

Introduction

Immunoglobulin G4-related disease (IgG4-RD) is characterized by high serum immunoglobulin G4 (IgG4) concentrations and pathologically IgG4-positive plasmacyte infiltrations and storiform fibrosis.[1)] Its etiology remains unknown. This disease affects multiple organs, either concurrently or metachronously. Steroid therapy is considered highly effective for disease control in IgG4-RD in nonvascular organs. Conversely, arterial lesions can lead to fatal outcomes due to rupture.[2)] No uniform consensus about the surgical indication for IgG4-related arterial disease has been achieved. We report a case of successful endovascular aneurysm repair (EVAR) of ruptured IgG4-related periaortitis.

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Case Report

An 80-year-old man underwent gastrectomy, pancreatic tumor enucleation, and right hemicolectomy at 70, 75, and 80 years of age, respectively. He was treated for chronic sialadenitis at 71 years of age. Serum IgG4 concentration increased to 693 mg/dl, and computed tomography (CT) revealed mild fibrous thickening of the soft tissue mass surrounding the abdominal aorta prior to pancreatic surgery. Although IgG4-RD had been suspected, thorough examination was not performed. The fibrous thickening of the soft tissue mass surrounding the abdominal aorta gradually grew since then. IgG4-related periaortitis was strongly suspected due to high serum IgG4 concentration of 1,760 mg/dl and conspicuous fibrous thickening of the soft tissue mass surrounding the abdominal aorta and the bilateral iliac arteries before the right hemicolectomy. CT scan revealed that the maximum short-axis diameter of the aorta, which was included in the fibrous thickening of the soft tissue mass, was 47 mm; it also revealed a pancreatic head mass 6 months prior to the present event (**Figs. 1A** and **1C**). The maximum short-axis diameter without the soft tissue mass was 25 mm (**Fig. 1D**). However, pathological investigation of IgG4-RD had not been conducted. He presented to our emergency department with sudden-onset abdominal pain and lumbago. On examination, he was alert, and his abdomen was bloated and generally tender. His systolic blood pressure was 81 mmHg and pulse rate 99 beats/min. After the massive fluid resuscitation, he remained hemodynamically stable and did not need blood transfusion. CT scan revealed a ruptured abdominal aorta (**Fig. 2**). We decided that endovascular repair was the appropriate treatment method due to the hostile abdomen. The patient underwent EVAR using a bifurcated stent graft (Gore Excluder aortic stent graft; W. L. Gore & Associates, Inc., Flagstaff, Arizona, USA). On intraopera-

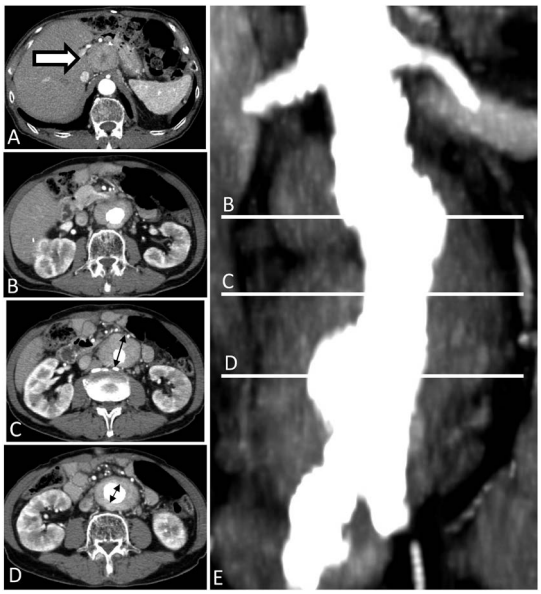

Fig. 1 Contrast-enhanced computed tomography (CECT) images 6 months prior to the rupture. (**A**–**D**) Sections of the axial CECT images. (**B**–**D**) The levels of the lines in (**E**). (**E**) Multiplanar reconstruction image.
(**A**) A pancreatic head mass (arrow). (**B**–**E**) Marked fibrous thickening of the soft tissue mass surrounding the abdominal aorta and extending beyond the aortic bifurcation. The maximum short-axis diameter of the aorta, which includes the soft tissue mass, is 47 mm at the level of line C (double arrow). The maximum short-axis diameter without the soft tissue mass is 25 mm at the level of line D (double arrow).

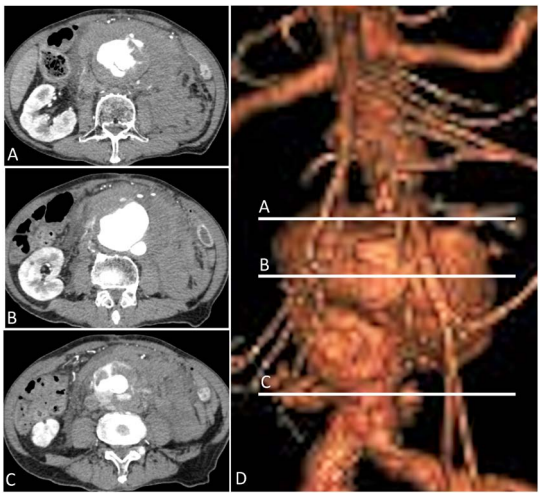

Fig. 2 Contrast-enhanced computed tomography (CECT) images at the time of the rupture. (**A**–**C**) Sections of the axial CECT scan images. (**A**–**C**) The levels of the lines in (**D**). (**D**) Three-dimensional CT image.
(**A**–**C**) Contrast medium entered the soft tissue mass surrounding the abdominal aorta. A huge hematoma can be observed in the retroperitoneal space. (**D**) Three-dimensional CT image of the abdominal aorta visualized as a complex form.

tive completion angiography, satisfactory deployment of the stent graft was confirmed. On the 7th postoperative day, a CT scan showed a satisfactory structural integrity/position of the patient's stent graft and no evidence of an endoleak. He was discharged 19 days later. Two years following EVAR, he began taking oral steroid therapy for the elevated serum IgG4 concentration of 1,440 mg/dl. One year following the initiation of steroid therapy, the fibrous thickening of the soft tissue mass and pancreatic head mass reduced (**Fig. 3**). The elevated serum IgG4 concentration also normalized.

Discussion

Since the first report of IgG4-RD in autoimmune pancreatitis in Japan in 2001,[3] IgG4-RD has been found in various systemic organs. Some inflammatory abdominal aortic aneurysms are diagnosed as IgG4-RD following pathological exmination.[4] Vascular surgeons must accurately diagnose inflammatory aneurysms as IgG4-RD or not. Either comprehensive or organ-specific diagnostic criteria are used to diagnose IgG4-RD in Japan.[5,6] Although our case had a soft tissue mass surrounding the abdominal aorta and bilateral iliac arteries, high serum IgG4 concentrations, a pancreatic head mass, a history of chronic sialadenitis, and the effect of steroid therapy, a definitive diagnosis of IgG4-RD could not be made using the aforementioned criteria. Pathological examination of the affected aorta will not be promising because endovascular treatment will play a major role in the aortic repair due to adherence to surrounding organs. To make a definitive diagnosis of IgG4-related arterial disease on the basis of the aforementioned criteria, pathological findings of vascular lesions or definitive diagnosis in nonvascular organs are essential. Thus, a definitive diagnosis of IgG4-RD will be difficult to make in the case of aortic involvement as the first manifestation.

When a diagnosis of IgG4-related arterial disease is made, assessment for the indication of treatment is needed, especially for dilated lesions. Steroid therapy was reported to be unsuitable for patients with preexisting luminal dilatation.[7] However, regardless of the existence of luminal dilatation, steroid therapy may be used according to the progression of IgG4-RD in nonvascular organs. Although aneurysmal dilatation of the aortic lumen was not present in our case, the rupture occurred prior to steroid therapy.

Many vascular surgeons have used the maximum short-axis diameter of the aorta as an indicator for surgical intervention of aneurysms. However, no consensus has been reached regarding the measurement of the diameter for IgG4-related aortic disease. Whether a soft tissue mass is included or not is uncertain. In our case, the maximum

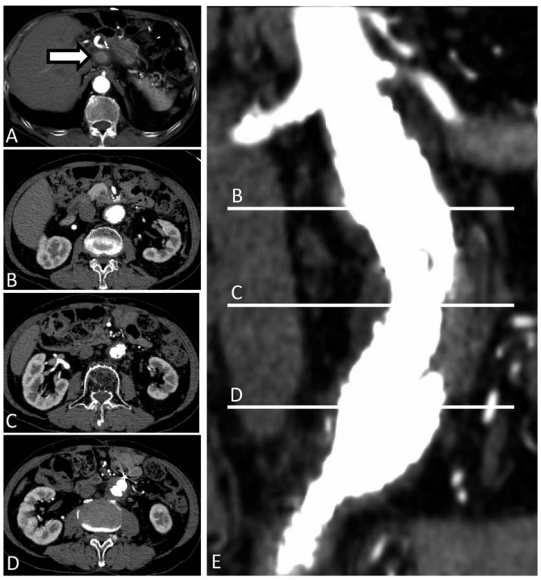

Fig. 3 Contrast-enhanced computed tomography (CECT) images 1 year following the initiation of steroid therapy. (**A**–**D**) Sections of the axial CECT scan images. (**B**–**D**) The levels of the lines in (**E**). (**E**) Multiplanar reconstruction image. The axial slice levels in (**A**) through (**D**) are about the same height as the axial slice levels in (**A**) through (**D**) in **Fig. 1**. (**A**) A pancreatic head mass reduced in size (arrow). (**B**–**E**) Reduced thickness of the soft tissue mass.

short-axis diameter of the aorta, which included a soft tissue mass, was 47 mm (**Fig. 1C**). The maximum short-axis diameter without the soft tissue mass was 25 mm (**Fig. 1D**). At both diameters, no indication for surgical intervention has been established for usual abdominal aortic aneurysms. We speculated that the aneurysmal wall in IgG4-related aortic disease was destructed by the expansion of the wall and the invasion of IgG4-positive cells to the intima.[8] Furthermore, we thought that some other novel indicator for surgical intervention is suitable for IgG4-related arterial disease. Tabata et al. reported that serial changes in serum IgG4 concentration in a patient with IgG4-RD were useful for evaluating disease activity, but not for the comparison of severity among patients with IgG4-RD.[9] In our case, serum IgG4 concentration greatly increased compared with that 6 years before rupture. Persistent elevation in serum IgG4 concentration may be one indicator. Zhang et al. reported that ^{18}F-fluorodeoxyglucose-positron emission tomography may also be effective for assessing disease activity.[10]

Conclusion

Ruptured IgG4-related periaortitis was demonstrated. We believe that the focus should not only be on the aortic diameter but also on disease activity for a timely surgical intervention. Further prospective studies on IgG4-related arterial disease should be conducted.

Disclosure Statement

The authors declare no conflicts of interest associated with this manuscript.

Additional Note

The patient has given permission to publish the features of his case, and we have ensured that his identity is protected.

Author Contributions

Study conception: MH
Data collection: MH
Analysis: MH
Investigation: MH
Writing: MH
Critical review and revision: all authors
Final approval of the article: all authors
Accountability for all aspects of the work: all authors

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