A 69-year-old man was referred to our department for surgical treatment of bilateral giant coronary artery aneurysms. He had no apparent clinical factors associated with atherosclerosis, such as diabetes mellitus, hyperlipidemia, or hypertension. Physical examination revealed no abnormalities. His medical history included malignant lymphoma that had been treated with chemotherapy and radiation therapy at the age of 60 years. He was also diagnosed with a plasma cell tumor in his tonsils at the age of 68 years. Just before treatment for this tumor, he developed sudden-onset myocardial infarction. Coronary angiography revealed 2 giant coronary aneurysms; one presented as a broad aneurysmal change of the right coronary artery (Figure 1A and Movie 1 in the online-only Data Supplement), and the other was located from the left main coronary artery to the left anterior descending artery (Figure 1B and Movie II in the online-only Data Supplement). On the basis of subsequent computed tomography, the maximum diameters of the right and left coronary aneurysms were 40 and 25 mm, respectively (Figure 2). Laboratory examination showed that the serum immunoglobulin G4 (IgG4) level was slightly elevated at 161 mg/dL (reference range, 4.8–105.0 mg/dL). As the extent of the myocardial infarction was considered to be rather small because of distal thromboembolism to a minor branch of the right coronary artery, conservative antiplatelet and anticoagulant therapies were administered, and no invasive treatment was undertaken at that time. Nine months later, after the plasma cell tumor had been clinically resolved with chemotherapy and radiation therapy, coronary angiography was performed again and disclosed that the aneurysms were further enlarged (Figure 1C and 1D and Movies III and IV in the online-only Data Supplement). Cardiac computed tomography measurement revealed that the maximum diameters of the right and left coronary aneurysms had increased to 53 and 40 mm, respectively (Figure 3A and 3B). Echocardiography also showed the bilateral coronary aneurysms with moderately deteriorated contraction of the left ventricle (Figure 4A and 4B). Despite the administration of prednisone, the serum IgG4 level significantly increased to 306 mg/dL.

Considering the risk of aneurysmal rupture and further thromboembolism, operative treatment was undertaken. The aneurysms were opened, and massive intraluminal thrombi were removed (Figure 5A and 5B). The entry and exit of both aneurysms and the orifices of their branches were closed, and quadruple coronary artery bypass grafting was performed. Histologic examination of the resected aneurysmal wall showed a massive lymphoplasmacytic infiltrate mainly in the adventitia (Figure 6A). Numerous IgG4-positive plasma cells were immunohistochemically identified (Figure 6B). The mean proportion of IgG4/total IgG-positive cells was 89.0%, which led to the diagnosis of IgG4-related periarteritis of the coronary arteries.

IgG4-related systemic disease was recently established as a novel clinicopathologic entity that can manifest in multiple organs, including the cardiovascular system. Inflammatory abdominal aneurysm has been considered to be a frequently occurring clinical condition; however, a few cases were recently reported in which pathologic changes recognized as IgG4-related disease were present in the coronary arteries. Although these previous reports described the appearance of tumorous change in the coronary arteries with remarkable thickness of the periarterial tissue, our patient exhibited extremely enlarged coronary aneurysms but without such a tumorous appearance. Steroid therapy is usually considered to be effective for IgG4-related disease. However, in our patient, the coronary aneurysm continuously expanded despite the administration of prednisone. This is similar to the case reported by Ikutomi et al, which also eventually required surgical resection of the aneurysm.

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Disclosures

None.

References


Figure 1. Coronary angiography at the onset of the myocardial infarction (A and B) and 9 months later (C and D). A, The right coronary artery was diffusely dilated. B, A huge aneurysm was seen on the left main coronary artery and the proximal portion of the left anterior descending artery (LAD). C, The right coronary artery became further enlarged, especially at its midportion. D, The aneurysm of the left coronary artery was also remarkably enlarged.
Figure 2. Volume-rendered image of the coronary arteries obtained from the first cardiac computed tomography examination. There was no significant stenosis; however, aneurysmal change was seen on the broad part of the right coronary artery and the proximal part of the left coronary artery.

Figure 3. Maximum-intensity projection image obtained by follow-up computed tomography. A, Coronal view demonstrated a huge aneurysmal lesion of the right coronary artery filled with massive intraluminal thrombi (arrow). B, Nearly axial image showed that the bilateral coronary aneurysms had remarkably expanded (arrows).
Figure 4. Transthoracic echocardiography at the follow-up examination. A, Parasternal short-axis image. Bilateral giant coronary aneurysms could be seen (arrows). B, Right-sided parasternal short-axis image. A huge right coronary aneurysm filled with massive intraluminal thrombi was clearly seen (arrow).

Figure 5. Intraoperative images. A, Huge right coronary aneurysm with glossy surface. B, The right coronary aneurysm was opened, and its internal thrombi were removed.

Figure 6. Histologic examination of the resected aneurysmal wall. A, Massive lymphoplasmacytic infiltration mainly in the adventitia and severe destruction of normal vascular structure were seen by hematoxylin–eosin staining. B, Immunohistologic analysis revealed numerous IgG4-positive plasma cells infiltrating the aneurysmal wall.
A Surgical Case of Expanding Bilateral Coronary Aneurysms Regarded as Immunoglobulin G4-Related Disease
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Movie Legend:

**Movie 1.** Right coronary artery image by coronary angiography (right anterior oblique view) at the onset of the myocardial infarction. Best viewed with Windows Media Player.

**Movie 2.** Left coronary artery image by coronary angiography (right anterior position) at the onset of the myocardial infarction. Best viewed with Windows Media Player.

**Movie 3.** Right coronary artery image by the follow-up angiography. Best viewed with Windows Media Player.

**Movie 4.** Left coronary artery image by the follow-up angiography. Best viewed with Windows Media Player.