Immunoglobulin G4 (IgG4)-related disease is rare. It is characterized by marked elevation in serum IgG4 concentration and infiltration of IgG4-positive plasma cells into a variety of tissues, particularly the adventitia surrounding great arteries. Here, we report the case of a patient who suffered an acute myocardial infarction and eventually died of a ruptured thoracic aorta as a result of IgG4-related disease.

An 84-year-old man was admitted to our hospital complaining of new-onset substernal compressing pain. The diagnosis of acute posterolateral myocardial infarction was made by means of ECG, ultrasound imaging, and the presence of elevated serum creatine phosphokinase and troponin-I levels. Subsequently, urgent coronary angiography showed a huge coronary aneurysm (21 mm at its maximal diameter) accompanied by thrombus in the left circumflex artery (Figure 1 and online-only Data Supplement Movie I). Insiginificant stenosis could be seen proximally in several branching arteries, and the right and left anterior descending arteries appeared patent (Figure 2A and online-only Data Supplement Movie II). Thrombus was successfully aspirated from the left circumflex artery, and a stent was subsequently implanted.

To further elucidate the anatomy of the left circumflex artery aneurysm, coronary computed tomography angiography (CCTA) was performed on the second day of admission. Notably, no obvious atherosclerotic changes or sclerotic obstructions were seen; however, most of the epicardial coronary arteries, including the left circumflex artery aneurysm, were diffusely and continuously surrounded by tumor-like lesions that looked like Pigs-in-a-Blanket Coronary Arteries named after a recipe in which sausages are wrapped in bacon or pastry (Figures 2B and 3 and online-only Data Supplement Movie III). A second coronary angiography was performed on the fifth day of admission so as to determine the relationship between the coronary endothelium and the adventitia. Intravascular ultrasound imaging in the right coronary artery clearly showed intact endothelium, but the adjacent tumor-like lesions exhibited a low echoic pattern that extended into the adventitia to depths of 3 to 6 mm (Figure 2C). Integrated positron emission tomography/computed tomography showed substantial accumulation of \(^{18}\)F-fluorodeoxygucose in the pericoronary tumor-like lesions and the submandibular glands. Accumulation of \(^{18}\)F-fluorodeoxygucose was also noted in the celiac artery and both renal arteries but not, at this stage, in the thoracic or abdominal aorta (Figure 4).
CCTA and positron emission tomography/computed tomography findings suggested a diagnosis of IgG4-related disease. Indeed, plasma IgG levels were found to be 8194 mg/dL (reference range, 870–1700 mg/dL), the majority of which was found to be IgG4 (measured as 2630 mg/dL, reference range <135 mg/dL). Treatment with an oral corticosteroid (prednisolone 30 mg/day), which is recommended as the initial therapy for IgG4-related disease,2 was initiated. His condition remained stable, so the dose was gradually tapered after 1 month. However, he unexpectedly died of a ruptured thoracic aorta 3 months after his initial presentation.

At autopsy, considerable irregular thickening was evident around all the epicardial coronary arteries (Figure 5A). Hematoxylin-eosin staining demonstrated aggregates of infiltrating lymphocytes and plasma cells, with deposition of hemosiderin in the adventitia. Immunostaining showed that the infiltrating plasma cells were IgG4 positive (Figure 5B and 5C). The site of the ruptured thoracic aorta also showed IgG4-positive plasma cell infiltration that we assume must have developed rapidly after the positron emission tomography/computed tomography imaging.

In this case, this rare disease was found by chance during CCTA imaging. To our knowledge, only a handful cases of IgG4-related coronary periarteritis have been reported.3,4 The CCTA imaging that revealed the Pigs-in-a-Blanket Coronary Arteries sign strongly suggested the diagnosis in this case. However, to establish the definitive diagnosis of this potentially devastating disease, detailed multimodal imaging is required so as to (1) differentiate the appearances from the subendothelial positive remodeling of the coronary arteries, rare vasculitides that affect large vessels (eg, Takayasu disease, Kawasaki disease), and tumors that involve coronary artery (eg, malignant lymphoma and Rosai-Dorfan disease); and (2) map the systemic distribution of IgG4-positive plasma cell infiltration that may extend beyond the coronary arteries.

Seeing the Pigs-in-a-Blanket Coronary Arteries on CCTA strongly suggests diagnosis of IgG4-related coronary periarteritis, but confirmation is required with biochemical and immunological tests. Subsequently, more widespread imaging is needed to detect extracardiac disease. Consideration should be given to surveillance imaging and measurement of serum IgG4 levels to ensure response to treatment.

Figure 2. Right coronary artery (RCA) images obtained by invasive coronary angiography, coronary computed tomography angiography (CCTA), and intravascular ultrasound (IVUS) imaging. A, RCA image (left anterior oblique view). There was no significant stenosis. See online-only Data Supplement Movie II. B, Curved multiplanar reconstruction image of RCA obtained by CCTA. The RCA also showed no significant atherosclerotic lesions. However, tumor-like lesions 8 to 10 mm in thickness from their proximal to distal portions are evident around the RCA (dotted arrows), named as the Pigs-in-a-Blanket Coronary Arteries. The computed tomography (CT) values of the tumor-like lesions were 37±7 Hounsfield Units (HU) in the nonenhanced CT images and 86±25 HU in the enhanced images (dotted arrows). C, IVUS image of the RCA. The lower image is the longitudinal RCA image obtained by an IVUS catheter. The upper image is the cross-sectional coronary image at the site of the white line (20 mm distal from the proximal RCA site). The diameter of the lumen was 3.1×3.2 mm (asterisk). There was a large low echoic lesion measuring 3 to 6 mm between the lumen and adventitia (dotted arrows). This lesion was surmised to be a tumor-like lesion on the CCTA.

Figure 3. Volume-rendered image of coronary arteries (coronary tree image) obtained by coronary computed tomography angiography. There was no significant stenosis in the right or left coronary arteries, but a huge coronary aneurysm can be seen in the left circumflex coronary artery (arrow). Tumor-like lesions were manually traced based on their computed tomography value (>50 Hounsfield units) on every axial image, colored green, and the images were combined with those of the coronary tree images.
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Disclosures

None.

References


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Yoji Urabe, Takashi Fujii, Shuji Kurushima, Shuji Tsujiyama and Yasuki Kihara

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