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). Insignificant 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 18F-fluorodeoxyglucose in the pericoronary tumor-like lesions and the submandibular glands. Accumulation of 18F-fluorodeoxyglucose 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, 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-Dorfman 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.
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Disclosures
None.

References

Key Words: IgG4-related disease • coronary computed tomography angiography • periarteritis • intravascular ultrasound • acute myocardial infarction

Figure 4. Integrated 18F-Fluorodeoxyglucose (FDG) positron emission tomography/computed tomography images. The FDG scan was performed after an 8-hour overnight fast to reduce the uptake of the radiopharmaceutical agent in the heart. Insulin was not required because this patient’s glucose levels were 128 mg/dL. In addition, he drank 500 mL of water to reduce the accumulation of excess radiopharmaceutical agent before FDG injection. The upper images are nonenhanced computed tomography images. The lower images are 18F-FDG positron emission tomography images at the same levels. Mild-to-moderate accumulation of 18F-FDG in tumor-like lesions, including the site of the aneurysm in the left circumflex coronary artery (arrow), right coronary artery (dotted arrow), and left anterior descending artery, was detected. In addition, small quantities of 18F-FDG can be seen to have accumulated in the celiac artery (asterisk), common iliac artery, spleen (double asterisk), and the parenchyma of both kidneys (arrow heads). However, there was no accumulation in the descending aorta.

Figure 5. Cross-sectional macroscopic image and immunohistochemical microscopic images of the right coronary artery (RCA). A, Macroscopic slice image of the proximal RCA. This image shows irregular thickening to depths of 3 to 8 mm and reddish brown coloring around the lumen (arrow). B, Low-power immunoglobulin G4 (IgG4)-immunostained microscopic image of the proximal RCA. The asterisk shows the arterial lumen. The adventitia has been infiltrated mostly by lymphocytes and IgG4-positive plasma cells. C, High-power view of IgG4-immunostained microscopic image, from the dotted square of B. Numerous IgG4-positive plasma cells can be seen in the adventitia.
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