Case Report

Mass lesions surrounding coronary artery associated with immunoglobulin G4-related disease

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Summary Immunoglobulin G4 (IgG4)-related diseases have been reported to be systemic diseases characterized by elevation of serum IgG4 concentration and infiltration of IgG4-positive plasma cells within the target organ. However, the involvement of coronary artery is very rare. Here, we report a 62-year-old man with mass lesions surrounding coronary artery and abdominal aorta associated with IgG4-related disease diagnosed by a needle biopsy of the mass lesion surrounding the coronary artery using echocardiography and computed tomography. After we started to treat the patient with prednisolone, his serum IgG4 level decreased, and the mass lesions of coronary and abdominal artery were markedly reduced in size after 4 months. In conclusion, IgG4-related disease should be considered in addition to tumors such as malignant lymphoma when mass lesions surrounding coronary artery are detected.

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Introduction

Immunoglobulin G4 (IgG4)-related diseases are systemic and are characterized by elevation of serum IgG4 concentration and infiltration of IgG4-positive plasma cells within the target organ. In particular, autoimmune pancreatitis is thought to be involved in IgG4-related sclerosing disease [1]. Other organs such as salivary gland [2], retroperitoneum [3], liver, bile duct [4], and lung [5] have been reported to be involved in IgG4-related disease. They are often sensitive to steroid therapy. The involvement of aorta in IgG4-related disease is relatively common and is often associated with retroperitoneal fibrosis. However, only several cases of the involvement of coronary artery have been reported so far [6–10]. We report a case of mass lesions surrounding coronary artery...
IgG4-Related disease of coronary artery

Figure 1  (A and B) Contrast material-enhanced computed tomography angiography shows the mass lesion surrounding the right coronary artery and abdominal aorta before corticosteroid therapy (arrowheads). (C and D) The mass lesion was markedly reduced in size after corticosteroid therapy (arrows).

associated with IgG4-related disease diagnosed by needle biopsy.

Case report

A 62-year-old asymptomatic man was admitted to our hospital for further examination because a cardiac tumor was incidentally found by echocardiography. He had received percutaneous balloon angioplasty in the right coronary artery (RCA) and left circumflex branch (LCx) of the left coronary artery (LCA) because of angina pectoris 18 years previously. In addition, he had undergone coronary bypass surgery [left internal thoracic artery to the left anterior descending branch (LAD) of LCA and saphenous vein grafts to RCA and LCx] 14 years previously. About 7 years ago, the saphenous vein grafts, RCA and LAD were all occluded, although the left internal thoracic artery graft was patent. Because fair collateral flow was observed from LCx to RCA, no further coronary intervention was performed and medical treatment was continued. He has now been treated with inhaled steroids for 3 years due to bronchial asthma.

His echocardiography showed normal left ventricular wall motion and a mass lesion in the atrioventricular sulcus, which compressed the right atrium. Thoracic computed tomography (CT) revealed a mass surrounding the RCA and proximal site of LAD (Fig. 1). Abdominal CT revealed a thickened infrarenal abdominal aortic wall (Fig. 1). An electrocardiogram showed small Q waves in the inferior leads.

On laboratory examination, C-reactive protein was 0.43 mg/dl (reference range, 0.0–0.17), total protein, 9.0 g/dl (6.7–8.3), albumin, 3.5 g/dl (4.0–5.0), and N-terminal pro-brain natriuretic peptide, 177 pg/ml (<131). His serum IgG was 3627 mg/dl (870–1700), IgG4, 2170 mg/dl (4.8–105), IgE, 681 IU/ml (<269), IgA, 168 mg/dl (110–410), IgM, 85 mg/dl (35–220), soluble interleukin-2 receptor 643 U/ml (145–518), and antinuclear antibodies were negative. IgG4 subtype made up 60% of the total IgG fraction (usually less than 5–6%).

Gallium-67 scintigraphy revealed no uptake into the mass lesion of coronary artery and abdominal aorta, and bone marrow aspiration showed no abnormal findings. Taken together, we suspected that the tumors surrounding the coronary and abdominal artery were associated with IgG4-related disease, although neither swollen pancreas nor significant retroperitoneal fibrosis was observed by abdominal CT. To confirm the diagnosis, we successfully performed a needle biopsy of the mass lesion surrounding the RCA via the epigastric fossa, using echocardiography and CT.

Histological assessment of the coronary mass lesion showed fibrous tissue with infiltration of lymphocytes and plasma cells. Many of the plasma cells were positive for
IgG and IgG4. The ratio of IgG4-positive cells/IgG-positive cells was > 60% and this indicated that the mass lesion was associated with IgG4-related disease (Fig. 2).

We then started to treat the patient with 40 mg (0.6 mg/kg) prednisolone daily to reduce the mass lesion and to prevent compression of the right atrium. As a result, his serum IgG4 level decreased to 219 mg/dl, and the mass lesions of coronary artery and abdominal aorta were markedly reduced in size 4 months. The dose of prednisolone was gradually tapered and was maintained at 7.5 mg without serious side effects. No recurrence was observed at medical follow-up of 1 year.

**Discussion**

The involvement of the aorta in IgG4-related disease is relatively common and is often associated with retroperitoneal fibrosis. However, the involvement of coronary artery is very rare. To the best of our knowledge, only 5 cases of IgG4-related tumorous lesions surrounding the coronary artery have been previously reported [6–10]. All 6 cases, including the present case, were reported in Japan. Characteristics of the 6 cases are summarized in Table 1. Five cases were elderly men, while one was an elderly woman. Four of the six cases had mass lesions surrounding both right and left coronary arteries, one surrounding RCA only, and one surrounding LCA only. Four cases had a coronary artery aneurysm; three of these had a thrombus and one had an acute myocardial infarction associated with an acute thrombus [2]. It was reported that IgG4-related periarteritis mainly occurred in the tunica externa and presented as an aneurysmal lesion rather than as stenosis [6].

At least four cases also had tumorous lesions surrounding the abdominal aorta. Two cases had other IgG4-related glandular diseases.

The diagnosis was confirmed by ultrasound and CT-guided needle biopsy in the present case, while the previous cases were diagnosed as follows: four cases by open chest biopsy and one indirectly by renal biopsy. We recommended ultrasound and CT-guided-needle biopsy because it minimizes the risk of coronary artery injury and cardiac tamponade. Moreover, it is less invasive compared with open chest biopsy; this method also has the advantage of permitting therapy to begin as soon as possible after diagnosis, in contrast to open biopsy.

IgG4-related disease is usually sensitive to steroid therapy. The present case and two previous cases were treated with oral prednisolone [7,8], and two cases were treated with surgical resection of the coronary aneurysm and/or abdominal aneurysm without steroid therapy [6,9], and one case was finally treated with surgical resection of the coronary aneurysm because steroid therapy was not fully effective for the coronary lesion [10].

Both of these therapies achieved a marked reduction in serum IgG4 levels, and the steroid treatment reduced mass lesion size surrounding coronary artery and aorta in 3 of 4 cases. One case died of aortic rupture after corticosteroid

![Image](https://example.com/image.png)
therapy. Although an association between rupture and corticosteroid therapy was previously unknown, we should treat patients with corticosteroids carefully because such therapy for IgG4-related periarthritis may induce thinning of the aortic wall.

In conclusion, IgG4-related disease should be considered in addition to tumors such as malignant lymphoma when mass lesions surrounding coronary artery are detected, and steroid therapy is effective for the coronary lesions in at least half of these patients.

References


