

A Case of Microscopic Polyangiitis with Giant Coronary Aneurysm

To the Editor:

Microscopic polyangiitis (MPA) is the most common myeloperoxidase-antineutrophil cytoplasmic antibody (MPO-ANCA)-associated small-vessel vasculitis, with few or no immune deposits in the involved vessels. MPA always affects small vessels (capillaries, venules, arterioles) but medium-size vessels can sometimes be involved^{1,2}. We describe a case of medium-size vessel MPA with a giant coronary aneurysm typically found in polyarteritis nodosa (PAN). To our knowledge, this is the first documented case of a giant coronary artery aneurysm in a patient with MPA.

A 78-year-old woman whose previous chief complaint was chest pain and who had no history of hypertension or dyslipidemia, smoking history, or any notable family history visited a hospital because of generalized malaise in 2005. Deterioration in renal function (Cr 2.82 mg/dl) was found, but a renal biopsy yielded no definitive diagnosis. Renal failure progressed (Cr 4.65 mg/dl) and a renal biopsy was repeated in 2006 because of high MPO-ANCA titer (258 EU). On the basis of the biopsy, crescentic nephritis (ANCA-associated nephritis) was diagnosed. However, steroid therapy was withheld because of advanced stage (endstage) of the disease (Cr was 4.73 mg/dl at discharge). Maintenance hemodialysis 3 times weekly was initiated in 2006. The patient continued with followup in spite of mild chest pain during walking that started in the summer of 2009. Due to aggravated malaise, low-grade fever, and subsequent frequent occurrences of pericardial tightness, she was referred to the Department of Cardiology of our hospital in August 2009. A coronary computed tomography (CT) angiography was performed because ischemic heart disease was suspected. This revealed a 14-mm diameter coronary artery aneurysm on the bifurcation of the left main trunk (Figure 1). Although there was low-grade fever and inflammatory findings (C-reactive protein 1.8 mg/dl, erythrocyte sedimen-

tation rate 124 mm/h), no definite infection was found. Subsequent coronary angiography revealed a saccular aneurysm on the bifurcation of the left main trunk and stenoses of 3 coronary vessels. After a coronary artery bypass graft was performed on the 3-vessel stenotic lesions in October 2009, chest pain improved but the low-grade fever and malaise remained. A chest CT scan in November 2009 revealed ground-glass opacity just under the bilateral lower lobe pleura. The patient was subsequently referred to the Department of Rheumatology. Based on persistent inflammatory findings, increased KL-6 and high MPO-ANCA titer (298 EU), a diagnosis of interstitial pneumonia associated with MPA was made. Given the age and history of the patient as well as the use of dialysis therapy, cardiovascular lesions were likely to be caused by arteriosclerosis. However, absence of hypertension and dyslipidemia, persistent inflammatory finding, high MPO-ANCA titer, and pulmonary manifestation suggested that cardiovascular lesions were possibly a complication of MPA. A covered stent graft was performed to seal the coronary artery aneurysm in January 2010, and prednisone 30 mg was started for treatment of MPA. Shortly after initiation of this therapy, MPO-ANCA titer began to decrease, along with improvement of both the inflammatory findings and pulmonary lesions.

In a series of 85 patients with MPA a surprisingly high frequency (50.6%) of cardiovascular manifestations was found. Pericarditis, heart failure, and hypertension were present in 9, 15, and 29 cases, respectively, and myocardial infarction in 23. The rare descriptions of cardiovascular complications of MPA could be explained by the likelihood that some of the earlier MPA patients had been misdiagnosed as having PAN. However, subclinical myocardial infarctions may be more common in MPA, as they are in PAN and other small-vessel vasculitides⁴. Coronary aneurysms are frequently seen in association with atherosclerosis, suggesting an overlap in risk factors and pathogenesis. It has been estimated that 50% of coronary aneurysms are due to atherosclerosis. The next most common cause is con-

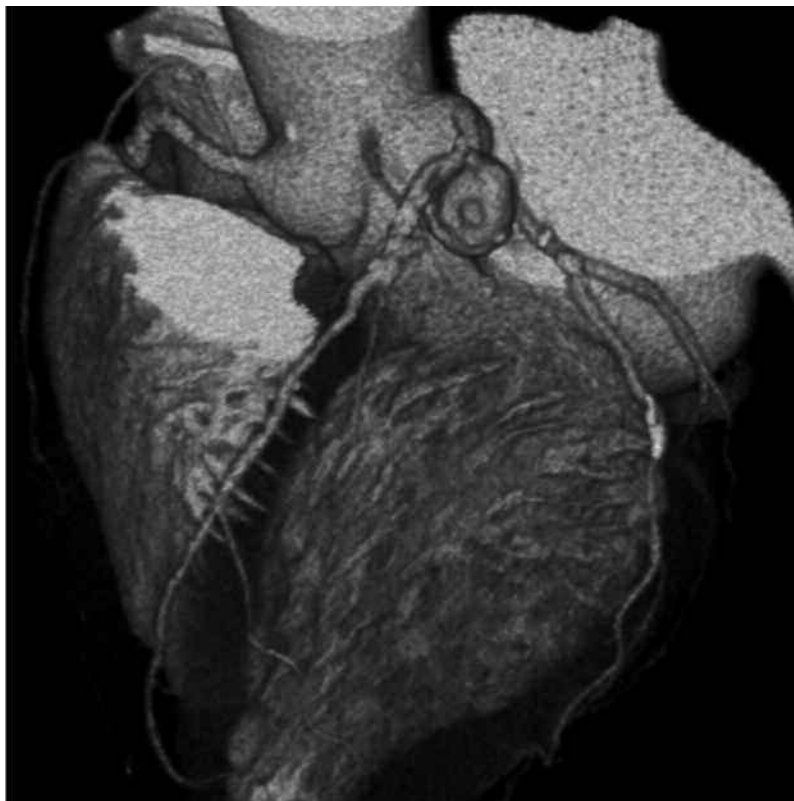


Figure 1. Coronary CT angiography shows a saccular coronary artery aneurysm at the left main coronary artery bifurcation.

genital, accounting for 20%–30% of coronary aneurysms. A host of inflammatory and connective tissue disorders have also been associated with coronary aneurysms. Most well known is the association with Kawasaki disease, but coronary aneurysms have also been reported in patients with Takayasu's arteritis, lupus, and rheumatoid arthritis⁵. However, aneurysm formation of medium-size and large vessels is a rare feature of MPA arteritis⁶. Our patient probably had ANCA-associated cardiovascular complication overlapped with pulmonary manifestation. Only one case of MPO-ANCA-associated glomerulonephritis complicated by an intrarenal aneurysm has been reported⁷. Aneurysms, mostly saccular, of the medium-size muscle arteries are frequently encountered in PAN. Disruption of the internal and external elastic lamina is noted and may contribute to the development of aneurysmal dilation⁸. In addition to arterial narrowing and thrombosis, inflammation can weaken the vessel wall and thereby lead to aneurysm formation.

The persistent inflammatory findings in this patient indicated that the cardiovascular lesion could not be explained by arteriosclerosis alone. Subsequent intercurrent occurrence of pulmonary manifestations suggested that coronary arteritis associated with vasculitis due to MPA caused formation of the coronary aneurysm.

This was a case of MPA associated with dialysis. Controlling the vasculitis was necessary to optimally improve this patient. There are few reports of aneurysm formation in patients with MPA, so this may be a very rare case.

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