

Coronary Artery Pseudoaneurysm due to Medial Mucoid Degeneration Mimicking an Intra-atrial Mass

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Abstract

Coronary artery aneurysms are frequently asymptomatic and may be difficult to diagnose by cardiac imaging. We herein present a case of a coronary artery aneurysm of the right coronary artery due to medial mucoid degeneration mimicking an intra-atrial mass on echocardiography and magnetic resonance imaging, with the cause being diagnosed after surgery.

Key words: coronary artery, aneurysm, cardiac mass, echocardiography, medial mucoid degeneration

(Intern Med 54: 2453-2458, 2015)

(DOI: 10.2169/internalmedicine.54.3804)

Introduction

Coronary artery aneurysms are defined as dilatations of the coronary artery with a diameter 1.5 times or more than that of the adjacent normal coronary artery (1). The incidence of coronary artery aneurysms was reportedly 1.5% to 5% in an autopsy series and 4.9% in the CASS registry (2, 3). It occurs most frequently in the distribution of the right coronary artery (RCA), followed by the left anterior descending coronary artery (LAD) and the left circumflex coronary artery (LCX), with men being more commonly affected (1, 2, 4). Although the aneurysm is often diagnosed by cardiac imaging studies in most cases, including coronary angiography (CAG), the diagnosis may be difficult. In two previous case reports, an accurate diagnosis could not be established until after surgery when the aneurysm presented as an intracardiac or myocardial mass (5, 6).

We herein present a case of a coronary artery aneurysm of the RCA which appeared as an intra-atrial mass on cardiac imaging (including echocardiography), CAG and cardiac magnetic resonance imaging (MRI), which was ultimately diagnosed by a histological study after the surgery.

Case Report

A 61-year-old man was referred to our institution after he was diagnosed with a cardiac tumor and aneurysm of the abdominal aorta by his family physician who he visited with the complaint of abdominal pain. He had been suffering from hypertension for the previous 30 years and had suffered a hemorrhagic stroke 8 years before this admission. Since then, he had right hemiparesis. A physical examination on admission revealed a blood pressure of 200/120 mmHg, pulse rate of 70 bpm and a palpable mass in the umbilical region of the abdomen. The left tibialis posterior and dorsalis pedis arteries were weakly palpable and his bilateral toes were cyanotic. Laboratory testing indicated a white blood cell count of 9,400/mm³, hemoglobin of 11.1 g/dL, blood urea nitrogen of 42 mg/dL, creatinine of 3.98 mg/dL, aspartate aminotransferase of 19 IU/L, alanine aminotransferase of 11 IU/L, lactate dehydrogenase of 316 IU/L, creatine kinase of 99 IU/L, C-reactive protein of 0.68 mg/dL, D-dimer of 17.3 µg/dL, and N-terminal pro-brain natriuretic peptide of 6,968 pg/mL. A chest X-ray showed mild cardiomegaly and normal lung fields. Electrocardiogra-

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Received for publication August 5, 2014; Accepted for publication February 12, 2015

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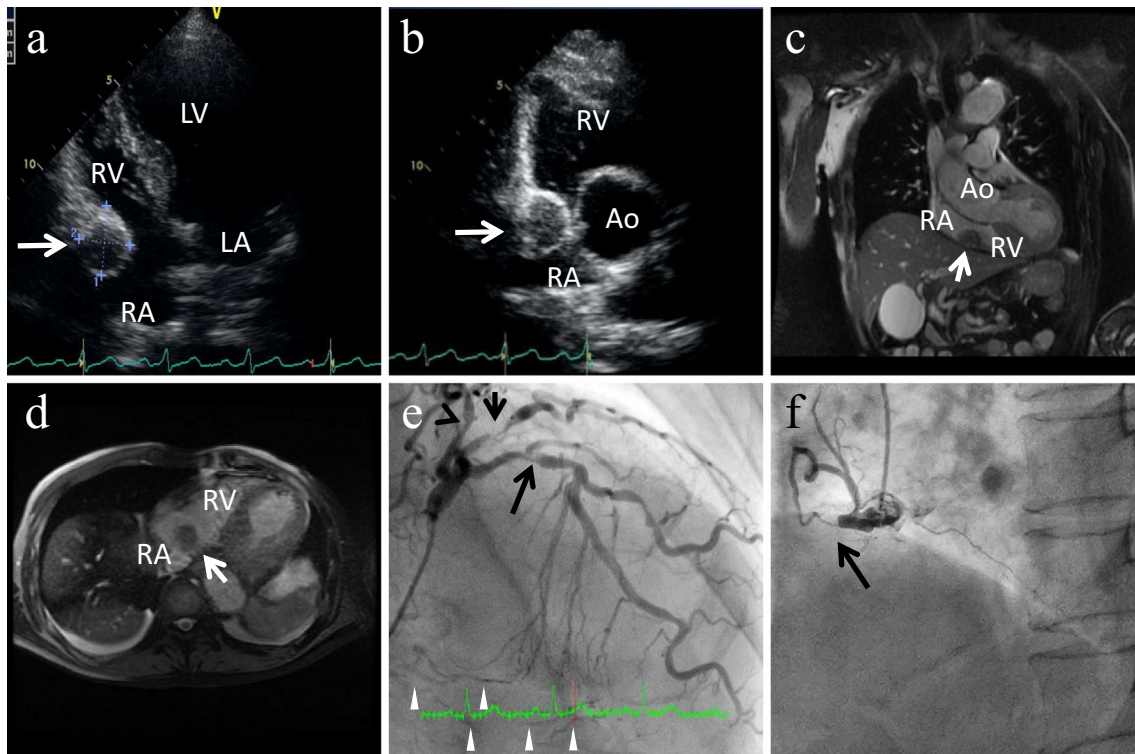


Figure 1. A round mass measuring 19×24×29 mm in size (arrows) is clearly visible near the anterior leaflet of the tricuspid valve in the right atrium via transthoracic echocardiography (a: the anteriorly angulated four-chamber view; b: the parasternal short axis view) and cardiac magnetic resonance imaging (T1 weighted image; c: coronal axis; d: horizontal axis) (Ao: aorta, RA: right atrium, RV: right ventricle). (e) Coronary angiography (CAG) of the left coronary artery (LCA) showed 99% stenosis in segment 6 of the left anterior descending artery (long arrow), 99% stenosis in segment 13 of the left circumflex artery (arrow head), and 99% stenosis in the high lateral branch (short arrow). Collateral circulation from the LAD and LCX was seen in the total occlusion of the RCA (white arrow heads). (f) CAG of the right coronary artery (RCA) indicated total occlusion in segment 1 of the RCA (arrow).

phy showed a first-degree atrioventricular block, left axis deviation and ST segment depression in V3-6. Transthoracic echocardiography (TTE) showed mild diffuse hypokinesis of the left ventricle (LV) with a LV ejection fraction of 55% and a round mass 19×24×29 mm in size near the anterior leaflet of the tricuspid valve in the right atrium (RA) (Fig. 1a, b). Cardiac MRI also demonstrated a mass in the RA (Fig. 1c, d). Chest and abdomen computed tomography (CT) showed an aneurysm of the abdominal aorta (infraarenal type, maximum diameter of 70 mm) in addition to aneurysms in the descending aorta (maximum diameter of 46 mm) and the common iliac arteries bilaterally (right, 25 mm in diameter; left, 20 mm in diameter). Enhanced CT was not performed due to the renal dysfunction.

After admission, the patient's blood pressure was controlled with the continuous intravenous infusion of nicardipine. CAG with a minimal dose of contrast medium was performed for the preoperative evaluation, which revealed a total occlusion of segment 1 of the RCA, 99% stenosis of segment 6 of the LAD, 99% stenosis of segment 11 of the LCX, and 99% stenosis in the high lateral branch of the left coronary artery. Collateral circulation from the

LAD and LCX was seen in the total occlusion of the RCA (Fig. 1e, f). We therefore opted to perform coronary bypass surgery and mass resection before surgery for the abdominal aortic aneurysm.

Intraoperatively, the mass was visible outside of the RA, but not in the right atrial cavity (Fig. 2a). The mass was excised from the outside of the right atrial wall (Fig. 2b). During the resection, two connections to the RCA were observed. However, there was no bleeding at resection due to the total occlusion of the proximal and distal parts of the RCA. After resection of the coronary aneurysm, the patient underwent coronary artery bypass grafting (left internal thoracic artery to the LAD and saphenous vein graft to segment 4 PD of the RCA). Approximately 5 weeks after the operation, Y-graft replacement surgery was performed for the abdominal aortic aneurysm.

A histological examination showed that the wall of the mass consisted of intima, media and adventitia, indicating that it was the coronary arterial wall, and the inside of the mass was a thrombus (Fig. 2c-e). The media of the coronary artery gradually thinned and disappeared, and some parts of the wall of the mass had only thickened adventitia, which

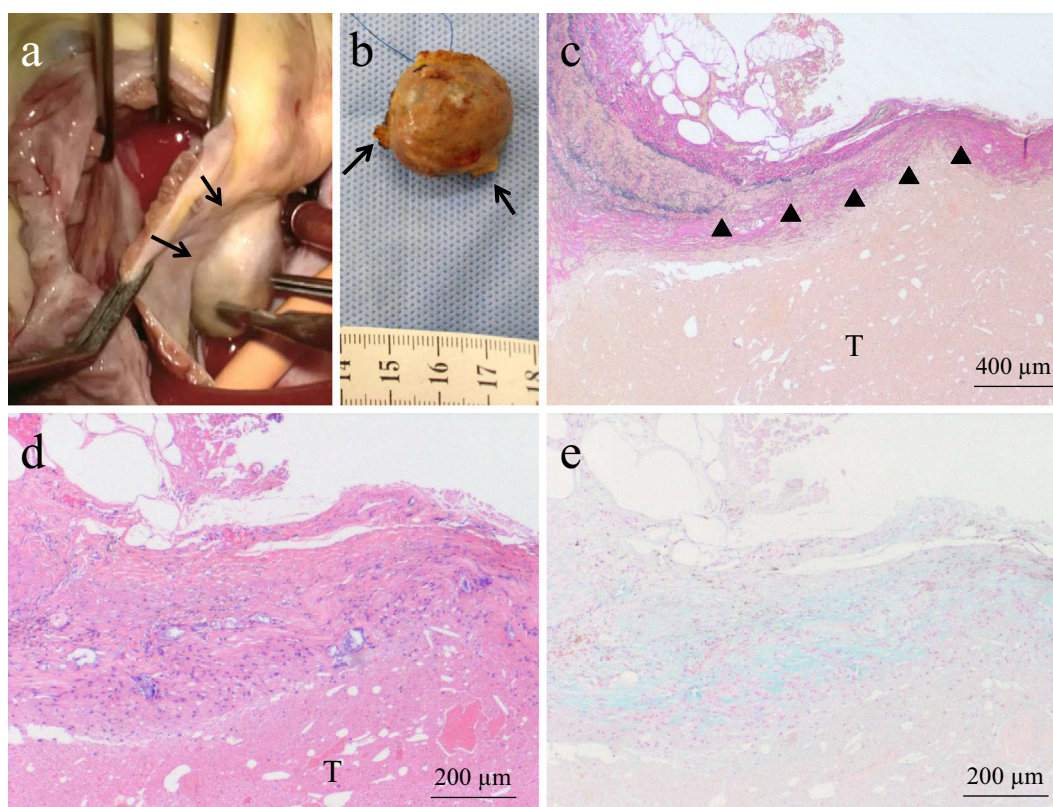


Figure 2. (a) After lifting the right atrial wall at surgery, the mass was seen outside the right atrium. (b) The extirpated round mass was 19×24×29 mm in size and was connected to the RCA (arrows). The wall of the mass indicated that it was the coronary arterial wall, and the inside of the mass was a thrombus (T). The thickness of the media of the coronary artery wall gradually decreased and some parts of the wall of the mass had only thickened adventitia (▲), indicating a pseudoaneurysm (c: Elastica van Gieson staining, magnification 20×). The coronary artery had medial degeneration, i.e., the disappearance of smooth muscle cells and interstitial deposition of Alcian blue-positive materials in the media (d: Hematoxylin and Eosin staining, magnification 40×; e: Alcian blue staining, magnification 40×).

was indicative of a pseudoaneurysm (Fig. 2c). Histologically, mucoid degeneration was visible in the media of the coronary artery (Fig. 2d, e). Therefore, the mass was diagnosed to be a pseudoaneurysm of the RCA due to medial mucoid degeneration with thrombus formation.

Unfortunately, the patient died of multiple organ failure approximately 1 month after the operation, and an autopsy was performed after receiving consent from his family. The histological examination of the abdominal aortic aneurysm indicated that it was a true aneurysm, and atherosclerosis and mucoid degeneration were seen in the tunica media of the abdominal aorta (Fig. 3).

Discussion

In the present patient, the coronary artery aneurysm was incidentally detected as an intracardiac tumor by echocardiography and MRI, although he had no chest symptoms related to the coronary aneurysm despite abdominal pain due to the giant aneurysm in the abdominal aorta.

Although most cases of coronary aneurysms are asymptomatic (7), some individuals may present with angina, myo-

cardial infarction (8) or syncope (9). An acute rupture in the cardiac chamber (10) or pericardial space causing tamponade (11), as well as fistula formation into the RA creating a left- to right-shunt (12), have been previously reported. These studies indicate that coronary artery aneurysms have no specific symptoms and signs.

Coronary artery aneurysms can occasionally be difficult to diagnose despite the availability of multiple cardiac imaging modalities, including CAG, MRI, TTE, and transesophageal echocardiography (TEE), and they may clinically present as cardiac (intracardiac and myocardial) or paracardiac masses (5, 6, 13-22). Thus far, ten cases of coronary artery aneurysms mimicking cardiac masses have been reported (Table) (5, 6, 13-18). In the present case, the aneurysm of the RCA in the atrioventricular groove presented between the RA and liver, compressing the right atrial wall against the cavity, making it appear that the mass was in the right atrium. In all the previously reported cases, the aneurysm was in the RCA or bypass graft to the RCA. In the previous cases, as well as in the present case, the maximum diameter of the aneurysm was 29 mm or greater. Taken together, the size of the RCA aneurysm appears to be related to its mim-

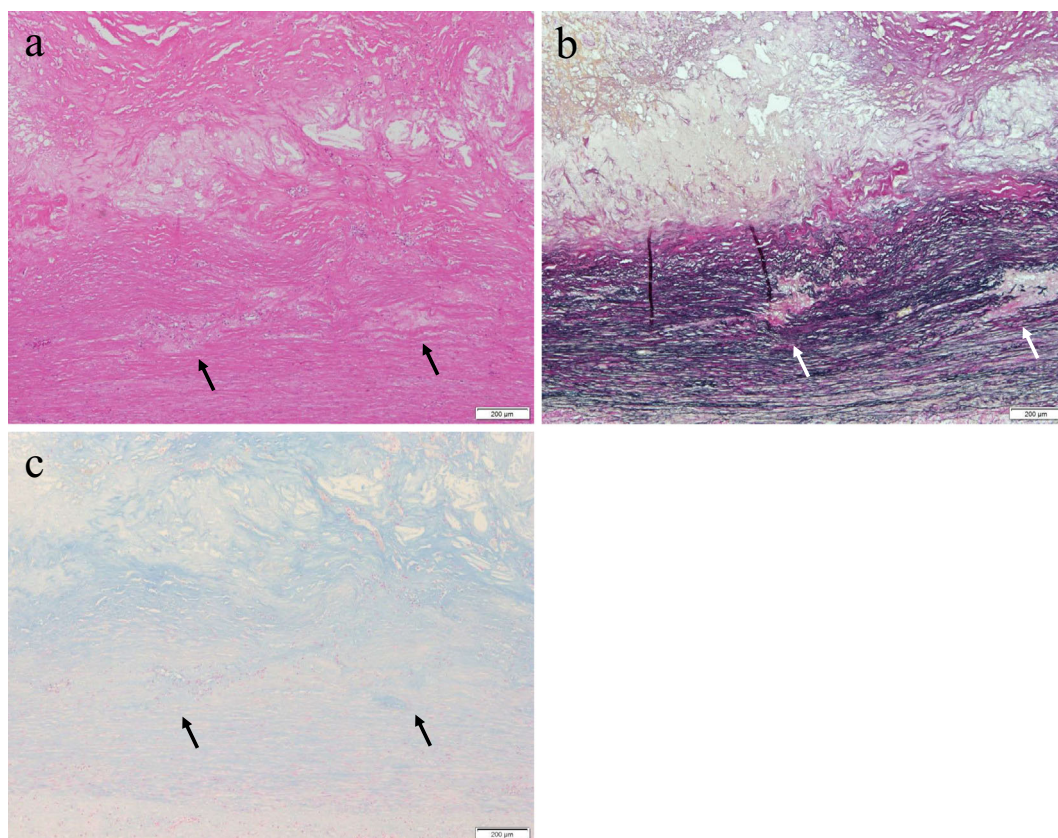


Figure 3. A histological examination of the autopsied abdominal aortic aneurysm showed that atherosclerosis was visible and mucoid degeneration (arrows) occurred in the tunica media of the abdominal aorta (a: Hematoxylin and Eosin staining, magnification 40×; b: Elastica van Gieson staining, magnification 40×; c: Alcian blue staining, magnification 40×; bar, 200 µm).

Table. Reports of Coronary Aneurysm Mimicking Cardiac Tumor or Intracardiac Mass.

Age (Ys)	Sex	Initial diagnosis	Location	Proximal total occlusion	Size (mm)	Histology	Ref
74	M	intraatrial mass	RCA	-	?	not exam	13
71	M	intracardiac mass	RCA	+	55×55	FMD	5
57	M	myocardial mass	RCA	-	50×50	not exam	14
73	M	cardiac tumor	RCA	+	50×60	atherosclerosis	15
60	M	cardiac tumor	RCA	-	30×30	atherosclerosis	15
55	M	cardiac tumor	RCA or graft	-	85×45×50	not exam	15
60	M	myocardial mass	RCA	+	52×61	not exam	6
46	?	cardiac tumor	RCA	+	75×60	atherosclerosis	16
49	M	intracardiac mass	RCA	-	50×45	not exam	17
38	F	intraatrial mass	RCA	-	74×71×80	atherosclerosis	18
61	M	intraatrial mass	RCA	+	19×24×29	mucoid degeneration	our case

Ys: years, M: male, F: female, RCA: right coronary artery, Proximal total occlusion: total occlusion of the proximal to coronary aneurysm, not exam: not examined, FMD: fibromuscular dysplasia, Ref: reference

icking a cardiac tumor.

In most previous cases, the aneurysm was diagnosed by CAG. However, in the present case, not even CAG was useful in the diagnosis because the RCA was totally occluded by a thrombus both proximal and distal to the aneurysm. The total occlusion proximal to the aneurysm was reported in five of the ten previous cases, and the diagnosis was not made until surgery in these cases, except for one case in

which CAG was not performed. Therefore, the total occlusion of the proximal side of the aneurysm appears to be related to the difficulty in the diagnosis of coronary artery aneurysms by CAG. However, three-dimensional CT CAG may be effective for the diagnosis if it is capable of being performed.

The most common cause of coronary aneurysms is atherosclerosis. Other causes include inflammatory processes

that affect the arterial wall directly, such as mucocutaneous lymph node syndrome (Kawasaki disease), Takayasu's disease, polyarteritis nodosa, systemic lupus, connective tissue disorders (e.g., Marfan or Ehlers-Danlos syndrome), septic emboli, syphilis and Lyme borreliosis. Less common causes include cardiac lymphoma, congenital coronary artery aneurysms, and trauma to the coronary arteries during angioplasty (16). Atherosclerosis is the main cause of coronary aneurysms mimicking cardiac tumors. In five of the ten previous cases who underwent a histological examination, the causes of coronary aneurysm were atherosclerosis in four and fibromuscular dysplasia in one (Table).

The present study showed that the cause of the pseudoaneurysm was medial mucoid degeneration of the coronary artery although the patient did not have Marfan syndrome. Pseudoaneurysms are less common than aneurysms of the coronary artery, and it has been previously reported in only one case of the reports of coronary aneurysms mimicking cardiac tumors or intracardiac masses, although that report did not mention the cause of the pseudoaneurysm (18).

Coronary pseudoaneurysm is mostly induced by catheter-based coronary intervention, and other causes include: spontaneous pseudoaneurysms, spontaneous dissection, blunt chest trauma or in association with cardiac tumors (23). There has only been one previous case report of a giant coronary artery aneurysm associated with medial mucoid degeneration, although whether it was a pseudoaneurysm or not was not mentioned (24). Therefore, to the best of our knowledge, the present case is the first to demonstrate that a coronary pseudoaneurysm may be induced by medial mucoid degeneration. Additionally, the present case demonstrated that there were several legions of mucoid degeneration in the tunica media of the abdominal aortic aneurysm associated with atherosclerosis. This suggests that the present patient may have had atypical Marfan syndrome in addition to atherosclerosis. Moreover, mild medial necrosis is one of the degenerative changes observed during aging, and a previous study demonstrated that chronic apoptosis of the vascular smooth muscle cells promoted medial degeneration in a murine model of atherosclerosis (25). Therefore, another possibility is that the combination of aging and severe atherosclerosis may enhance the progression of medial mucoid degeneration in the present patient.

In conclusion, coronary artery aneurysms should thus be considered in the differential diagnosis of intra-atrial masses, even when they are not visible on CAG due to total arterial occlusion, although the differential diagnosis of a cardiac mass typically only includes thrombus, tumors (primary or metastatic) and pericardial cysts.

The authors state that they have no Conflict of Interest (COI).

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