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Giant Right Coronary Artery Aneurysm Mimicking a Mediastinal Cyst With Compression Effects: A Case Report

Heidar Dadkhah Tirani,[1] Manouchehr Aghajanzadeh,[1] Reza Pourbahador,[2] Rasool Hassanzadeh,[3,*] and Hannan Ebrahimi[1]

[1]Department of Cardiothoracic Surgery, Guilan University of Medical Sciences, Rasht, IR Iran
[2]Department of Cardiology, Guilan University of Medical Sciences, Rasht, IR Iran
[3]Student Research Committee, Guilan University of Medical Sciences, Rasht, IR Iran

[*]*Corresponding author*: Rasool Hassanzadeh, Student Research Committee, Guilan University of Medical Sciences, Rasht, IR Iran. Tel: +98-9112335331, Fax: +98-1316690036, E-mail: rasool.hassanzadeh@gmail.com

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Abstract

Introduction: Giant coronary artery aneurysm is an extremely rare form of coronary artery disease. The most common cause of coronary artery aneurysms is atherosclerosis. Although it is usually asymptomatic, it may have various clinical presentations, including angina, myocardial infarction or sudden death.
Case Presentation: A 32-year-old woman presented with edema of the upper and lower limbs, palpitation, and chest pain, and was diagnosed with a giant right coronary artery aneurysm that had initially mimicked a mediastinal cyst. Although computed tomography (CT) suggested a mediastinal cyst, trans-thoracic echocardiography revealed an extra pericardial cyst. The definitive diagnosis of right coronary artery aneurysm was made based on CT angiography and coronary angiography findings. As treatment, aneurysmectomy was performed, and she was discharged on the sixth postoperative day with good general health condition.
Conclusions: Coronary artery aneurysm should be a differential diagnosis in cases of mediastinal cyst and mass lesion.

Keywords: Mediastinal Cyst, Echocardiography, Coronary Aneurysm, Coronary Angiography

1. Introduction

Small coronary artery aneurysms are seen in up to 5% of patients undergoing coronary artery angiography (1). Huge aneurysms that cause a remarkable bulge of the heart contour are rare and may mimic a mediastinal mass or cyst (2, 3). Atherosclerosis, as the most common cause of these aneurysms, is often due to other atheromas in the coronary arterial tree and is associated with poststenotic dilatation and ectasia (1). Another leading cause is Kawasaki disease, which is usually detected in infants and children but a coronary artery aneurysm may surface much later in adulthood (4). Coronary artery aneurysms are usually asymptomatic (5), but may present with clinical features such as angina, myocardial infarction, or sudden death (6). There could also be an acute aneurysmal rupture into a cardiac chamber or pericardium (1). Fistula formation into the right chamber is however rare (1). The aneurysm may also have compression effects on the surrounding structures (6). Herein, we report a case of a huge coronary artery aneurysm with compression effects on the superior and inferior vena cava and right ventricle that had clinically manifested as upper and lower limbs edema and mimicked a mediastinal cyst on imaging evaluation.

2. Case Presentation

A 32-year-old woman was referred to our department with a 5-month history of exertional dyspnea and a 2-week history of intermittent palpitation. She had consulted the emergency department of a local hospital for palpitations 4 weeks ago, but the palpitations subsided when she reached the hospital. During the next 2 weeks, her dyspnea increased and was almost persistent. She experienced fullness in the chest, frequent palpitations, and edema in the ankle and face, which was treated with hydrochlorothiazide. She was later referred to a cardiologist for further management. An echocardiogram revealed the presence of an extrapericardial large cystic lesion markedly compressing the right side of the heart. Computed tomography (CT) scan revealed a large heterogeneous cystic mass in the right anterior mediastinum (Figure 1A).

The patient was referred to a subspecialty hospital for surgical management. A cardiologist examined her again. A second echocardiogram showed the same results as described above. Contrast-enhanced CT scan revealed a large, heterogeneous, enhancing, cystic mass in the right anterior mediastinum; it had a maximum diameter of 97 mm and showed peripheral calcifications (Figure 1B). Based on

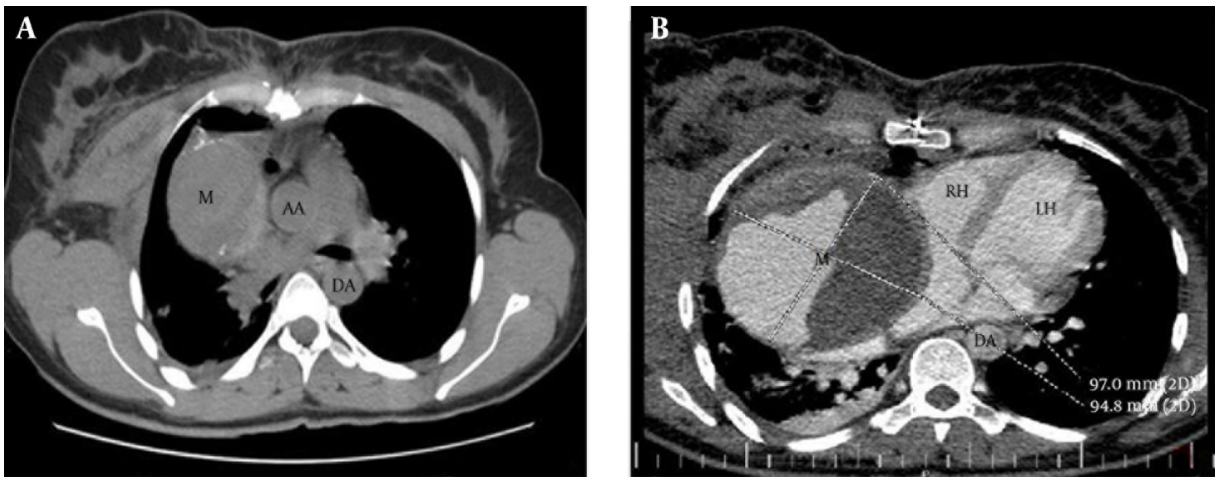

Figure 1. A, A CT image showing a large middle mediastinal mass with peripheral calcifications (M = mass, AA = ascending aorta, DA = descending aorta); B, enhanced CT image showing a large inhomogeneous, enhancing (97 mm), middle mediastinal mass with solid and cystic components compressing the right side of the heart. M = mass, RH = right side of the heart, LH = left side of the heart, DA = descending aorta.

echocardiography and CT scan findings, differential diagnoses included hydatid cyst, mediastinal mass, and cystic lesion and dermoid cyst. With the diagnosis of mediastinal cyst or mass, surgical exploration (medial sternotomy) was performed. A huge cystic mass with a thick wall was noted. Its right margin was free but the left margin was adhered to the right atrium and ventricle and compressed these structures. Fresh red blood was aspirated. Other differential diagnoses that surfaced included aneurysm of the Valsalva sinus, giant right coronary artery aneurysm, pseudoaneurysm of the aortic sinus, hemangiosarcoma, and cardiac tumor.

The cardiologist and cardiac surgeon recommended further evaluation by CT angiography and coronary angiography. The mid sternotomy was closed. CT angiogram revealed a large aneurysm in the distal portion of the right coronary artery with an intact proximal portion (Figure 2A). Coronary angiography also showed the same results (Figure 2B).

CT angiography and coronary artery angiography provided the definite diagnosis of right coronary artery aneurysm. Under general anesthesia and by mid sternotomy, cardiopulmonary bypass was initiated by cannulation of two veins, namely, the right femoral vein and the innominate vein. Antegrade cold blood cardioplegia was infused in the aortic root. The right coronary artery was exposed and snared by silk. Next, the hard wall of the aneurysm was opened; substantial blood clot and debris were removed; and most of the aneurysm wall was resected. The distal part of the right coronary artery could not be identified. Absence of any communication to the cardiac cavity was confirmed. Last, the right coronary artery was ligated by a Prolene stitch. The patient was eventually weaned off the cardiopulmonary bypass without any complication. The rheumatology work-up that followed was unremarkable. The patient was discharged at the sixth postoperative day under good general condition. During the 2-year follow-up period, the patient has had no complaints.

3. Discussion

A coronary artery aneurysm is defined as a focal dilation of the coronary artery to more than 1.5 times its normal size (1). Huge coronary artery aneurysms are rare. The incidence of coronary artery aneurysm is about 1.5% to 5%, and its etiology includes conditions such as atherosclerosis, congenital abnormalities, Kawasaki disease (7), connective tissue diseases, tumors, infections, iatrogenic and traumatic causes (8), systemic lupus erythematous, polyarteritis nodosa, Marfan's syndrome, Takayasu's arteritis, Ehlers-Danlos syndrome, Lyme borreliosis, septic emboli, syphilis, and primary cardiac lymphoma (7, 9, 10). Coronary artery aneurysms can also develop after procedures such as intracoronary Cook stent implantation, percutaneous coronary intervention, and directional coronary atherectomy. Blunt traumatic chest injury is another known cause. Coronary artery aneurysms are also associated with other diseases such as coronary artery angiodysplasia and idiopathic hypereosinophilic syndrome (8). Coronary aneurysms occur in 9% to 15% of children with Kawasaki disease (10). Our patient, however, had ev-

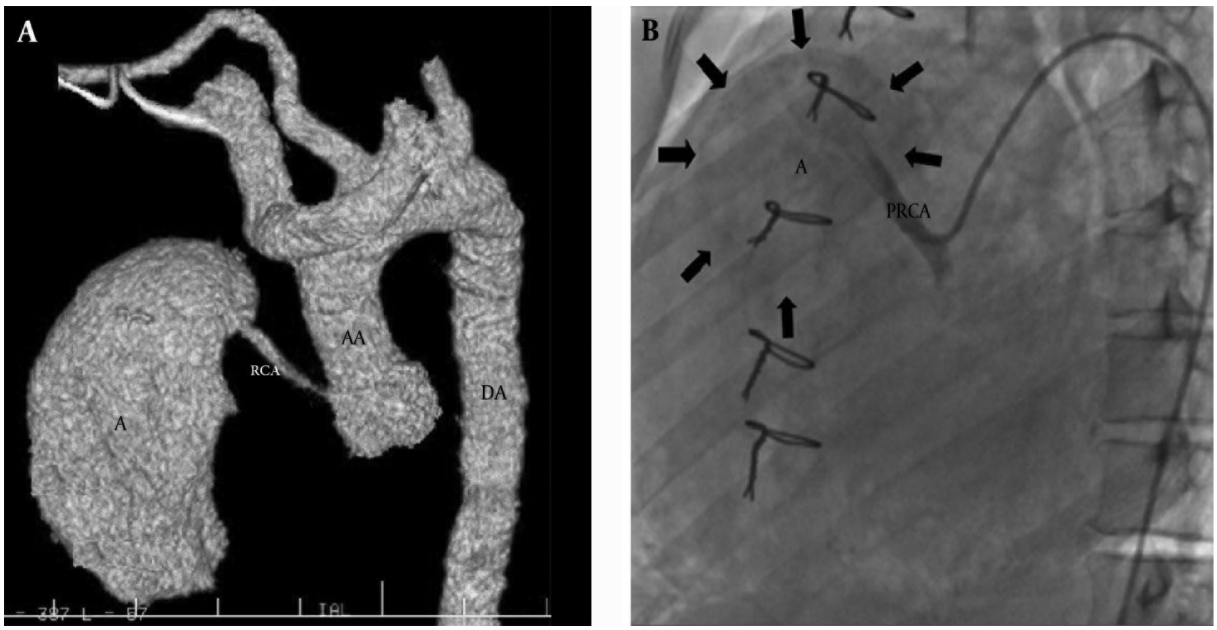

Figure 2. A, CT angiography image showing a large aneurysm in the distal portion of the right coronary artery. The proximal portion is intact. AA = ascending aorta, DA = descending aorta, RCA = right coronary artery, A = aneurysm; B, coronary artery angiography image showing a huge right coronary artery aneurysm. The proximal portion of the right coronary artery is intact. PRCA = proximal part of the right coronary artery, A = aneurysm.

idence of neither generalized atheromatous disease nor other possible causes of coronary artery aneurysm.

Patients with coronary artery aneurysms frequently present with dyspnea, angina pectoris, or sudden death. Based on these clinical presentations, it is difficult to differentiate coronary artery aneurysms from conditions such as coronary atherosclerosis, coronary vasculitis, coronary stenosis, coronary thrombosis, and acute or old myocardial infarction (2-7). Our patient had presented with palpitation and generalized edema.

Aneurysm of the heart wall, posttraumatic pseudo aneurysm of the ascending aorta or the pulmonary trunk, tumor of the heart or pericardium, and thymoma are considered the differential diagnoses of giant coronary aneurysm (7).

When the contrast material is injected, the turbulence of enhanced blood in a large aneurysm may simulate a heterogeneous enhancing mass, as was noted in our case. Moreover, in our patient, the peripheral linear calcifications and the typical location of the mass suggested a mediastinal cystic mass. Non-invasive imaging modalities like echocardiography and CT, although suggested the diagnosis, were not confirmative (5).

The exact diagnostic tool in such a case is the angiogram. Three-phase CT angiography helps in arriving at a diagnosis by showing homogeneous and similar densities of the mass and cardiac chambers in the unenhanced and equilibrium phases, and turbulence-like enhancement in the arterial phase. Radiologists should therefore be familiar with these CT angiographic characteristics, because a false interpretation of a soft-tissue mediastinal mass may prod biopsy examination, which may lead to a fatal outcome (11).

Magnetic resonance imaging (MRI) helps diagnose a giant coronary aneurysm, but it is unable to show the typical linear peripheral calcifications of the aneurysms. CT angiography not only produces two- and three-dimensional reformations and helps cardiac surgeons in preoperative planning by clearly depicting the spatial relations between the huge aneurysm and surrounding structures including the great vessels and the heart, but is also faster, more accessible, and cheaper than MRI in many medical centers (11). In the case described herein, surgical exploration confirmed that the sac was a true right coronary artery aneurysm that communicated proximally with the right coronary artery ostium and its distal communication remained identified and ended in the aneurysm sac. During the operation, the right coronary artery was ligated proximally and the aneurismal wall was maximally excised.

There is no consensus on the optimal management strategy for coronary artery aneurysms. The use of distal embolization and the effects of antiplatelet and anticoagu-

lant agents have been reported. Patient's management depends on the location of the aneurysm and the clinical context (2). Coil embolization and stenting have been used as non-surgical management of coronary artery aneurysms (12). Some surgeons believe that surgical repair is required when a coronary aneurysm is three times larger than the normal one (2, 12). Regarding coronary artery aneurysm being a very rare condition, there is no large randomized clinical trial that compares different therapeutic management approaches; consequently, the knowledge of optimal management is based on case reports, small case series, and personal experiences (2, 9, 12). In our patient, the right coronary artery aneurysm wall was opened via a mid sternotomy and cardiopulmonary bypass, substantial blood clot and debris were removed, and most of the aneurysm wall was resected. Although the prognosis of coronary artery aneurysm is controversial, the overall 5-year survival is reported to be 71% (1). Our patient was in good general condition during the follow-up period.

In conclusion, giant coronary artery aneurysms are uncommon lesions with various clinical presentations and should be included in the differential diagnosis of other conditions with similar clinical presentations. For the diagnosis and optimal management of these rare abnormalities and associated lesions, appropriate imaging techniques must be used. Surgical management should be planned carefully, as use of proper techniques would lead to better outcomes. Several cases of coronary artery aneurysms have been previously reported, but our case is rare and interesting because of its uncommon presentation mimicking a mediastinal mass accompanied with edema of the upper and lower limbs.

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Footnote

Authors' Contribution: Study concept and design and management of the case: Heidar Dadkhah Tirani; study supervision and surgical procedure: Manouchehr Aghajanzadeh; the referring and treating cardiologist: Reza Pourbahador; collection of the data, critical revision of the manuscript for important intellectual content, and guarantor: Rasool Hassanzadeh; drafting of the manuscript: Hannan Ebrahimi.

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