

An Unusual Huge Coronary Artery Aneurysm with Fistula

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Coronary artery aneurysms are noted in 0.15% to 4.9% of patients undergoing coronary angiography. Atherosclerosis accounts for 50% of coronary aneurysms in adults.¹ We report a 64-year-old female with a huge right coronary artery aneurysm with fistula connecting the aneurysm with the pulmonary artery and a fistula connecting the proximal left anterior descending artery with the pulmonary artery. Surgical resection of the coronary artery aneurysm and suture ligation of the coronary artery fistulas were performed. Pathological examination disclosed aneurysm with focal fibrosis and calcification. The patient presenting shortness of breath and angina-like symptoms caused by a huge right coronary artery aneurysm with compression of right ventricular outflow tract made this case noteworthy. The patient's symptoms resolved after surgical intervention.

Key Words: Coronary artery aneurysm • Coronary atherosclerosis • Coronary fistula

INTRODUCTION

Aneurysmal coronary artery disease is thought to occur in less than 5% of patients. Reported incidence rates vary, depending on the angiographic criteria used to define the aneurysms. Coronary artery aneurysm was defined as coronary dilatation that exceeds the diameter of normal adjacent segments or the diameter of the patient's largest coronary vessel by 150%.¹ The most common cause of coronary artery aneurysm in adult patients is atherosclerosis. Other causes include Kawasaki's disease, diagnostic or interventional coronary angiography, inflammatory and infectious arteritis, connective tissue

disease, syphilis, aortic dissection, tumor metastasis, trauma, and congenital malformation. Coronary artery fistula is an infrequent anomaly of coronary artery, and it may result in myocardial ischemia, congestive heart failure, and endocarditis.² We report an unusual case of a huge right coronary artery aneurysm with fistula connecting the aneurysm with the pulmonary artery.

CASE REPORT

A 64-year-old female had suffered from effort-related chest tightness and shortness of breath for one year. The chest tightness radiated to the left scapular region occasionally and lasted more than half an hour. However, the symptom could not be relieved by sublingual nitroglycerin. The patient had history of hypertensive cardiovascular disease for 2 years, hyperlipidemia for 2 years and type 2 diabetes mellitus with oral hypoglycemic agents therapy for one year. Chest radiography showed widening of the upper mediastinum (Figure 1). She received thallium-201 myocardial perfusion scanning that showed normal results, and chest computed tomography (CT) revealed a well-defined and well-enhanced lesion

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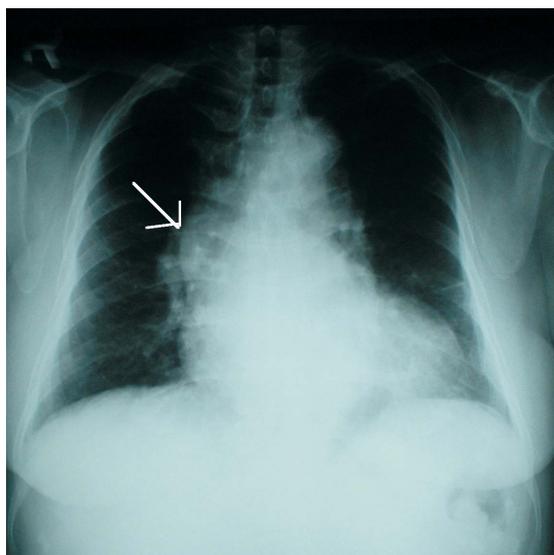


Figure 1. Chest radiography showed widening of the upper mediastinum (arrow).

about 5 cm in size in the right atrioventricular groove with suspicion of coronary artery aneurysm. Magnetic resonance imaging (MRI) study of the heart and magnetic resonance angiography of the aorta & pulmonary artery showed a well-defined mass about 5 cm in size in the right atrioventricular groove of the heart with the same signal intensity with connection to the right coronary artery and mild compression of the right ventricular outflow tract (Figure 2). Right coronary artery (RCA) aneurysm was impressed. Coronary angiography demonstrated no significant stenosis of coronary arteries, but a huge RCA aneurysm about 5 cm in diameter originating from the ostium of RCA with faint blood flow to the pulmonary artery (PA) suggesting RCA aneurysm with fistula connecting the aneurysm with PA (Figure 3). There was also a fistula connecting the proximal left anterior descending artery (LAD) with the PA (Figure 4). The patient underwent surgical intervention with aneurysmectomy of the RCA and suture ligation of the fistulas. Pathological examination revealed aneurysm with focal fibrosis and calcification. The patient's symptoms of chest tightness and shortness of breath resolved after operation. She was discharged in stable condition.

DISCUSSION

Coronary artery aneurysms were first recognized in

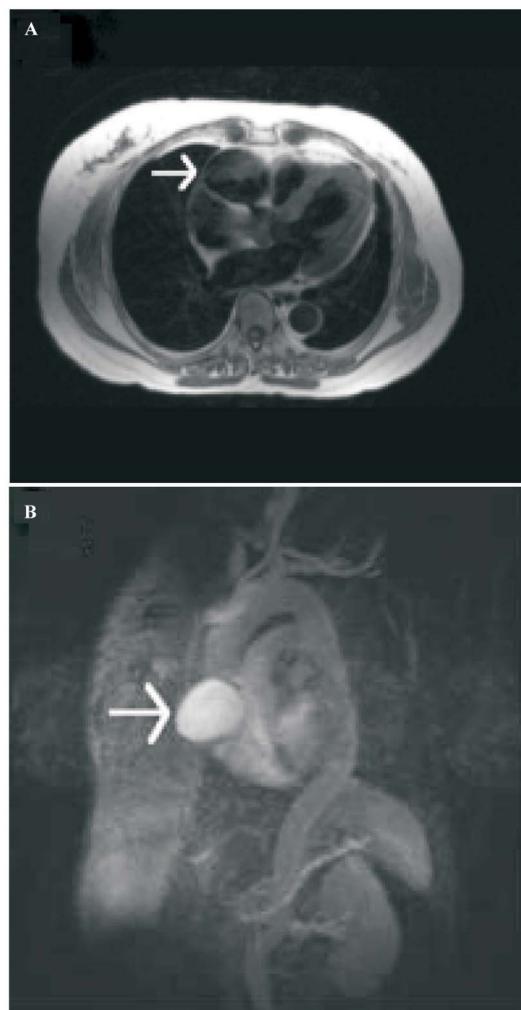


Figure 2. Magnetic resonance imaging demonstrated a large coronary aneurysm, measuring 5 cm in diameter, originating from the very proximal segment of the right coronary artery with compression to the right ventricle (A) and right ventricular outflow tract (B).

postmortem studies. With the advent of selective coronary arteriography, coronary aneurysms have been diagnosed with increased frequency. They have been reported in 0.15-4.9% of patients with suspected coronary artery disease, with a pronounced male predominance. Coronary artery aneurysms are most commonly found in the proximal and mid-portions of the RCA and, to a lesser extent, in the proximal portion of the left anterior descending artery or left circumflex artery. Atherosclerosis is common in western populations, and it has been hypothesized that aneurysmal dilatation may be a variant of atherosclerotic coronary artery disease.³

The long-term prognosis and optimal management

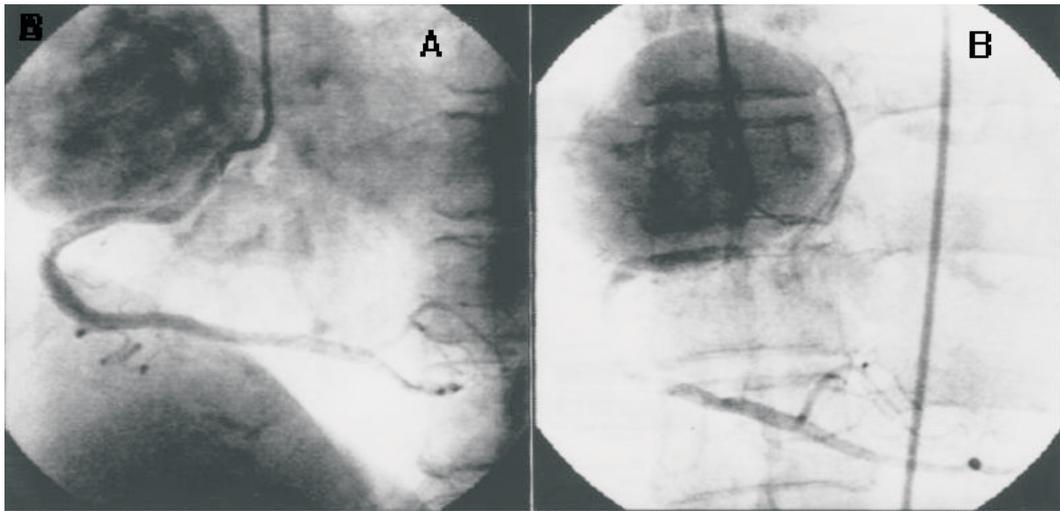


Figure 3. Coronary angiogram showed a huge right coronary artery (RCA) aneurysm, 5 cm in diameter with calcified wall, originating from the ostium of RCA and blood flow to the pulmonary artery via the fistula. (A) Left anterior oblique view. (B) Anterior-posterior cranial view.

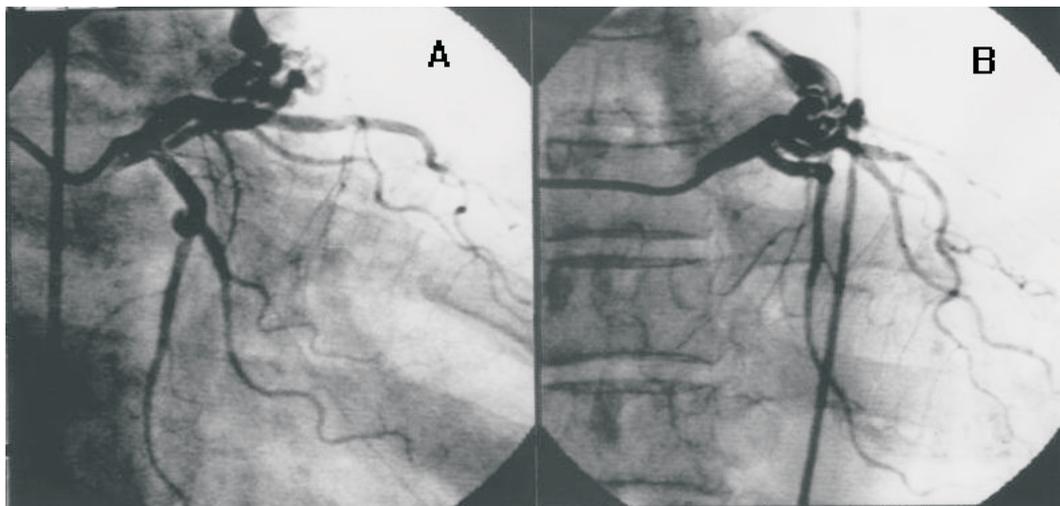


Figure 4. Coronary arteriovenous fistula originating from the left anterior descending artery with drainage into the pulmonary artery. (A) Right anterior oblique caudal view. (B) Anterioposterior view.

of these patients are uncertain. The altered blood flow in aneurysmal segments is thought to predispose to thrombosis or embolism and subsequent ischemia or infarction.^{3,4} Several reports support the notion that thrombosis and embolism do occur in patients with coronary aneurysms, but the clinical implications of this complication remain unclear.⁵ The pathophysiologic mechanisms of coronary artery aneurysm or ectasia in atherosclerotic coronary disease are probably multiple and involve the destruction of the vessel media, which results in dilatation of artery segments. Hence, turbulent flow in the area of expansion may contribute to further

dilatation. Histologic study of atherosclerotic coronary aneurysms reveals diffuse hyalinization, lipid deposition, focal calcification, fibrosis, and disruption of the intima with extension into the media and intramural hemorrhage.^{6,7} Traumatic injury of the coronary vasculature by coronary intervention device was another etiology of coronary aneurysm. The prognosis of patients with atherosclerotic coronary aneurysms has been shown to be similar to that of patients with obstructive coronary disease and seems to be related to the degree of aneurysmal dilatation.³

Hirose et al. showed that the combination of coro-

nary artery aneurysm and coronary artery fistula (coronary artery aneurysm associated with fistula, CAAAF) is extremely rare, and only 50 cases have been reported.⁸ CAAAF occurred more in females than in males (ratio = 2.2:1). One-third of the patients with CAAAF were asymptomatic, and two-third of patients may experience the symptoms of chest tightness⁹ or congestive heart failure such as dyspnea on exertion or palpitation. CAAAF may present with cardiac murmur or abnormalities on chest radiography.⁸ Aneurysm repair, fistulous closure and coronary artery bypass grafts are definitive treatments for CAAAF.⁸ We have presented this case who was diagnosed with a huge RCA aneurysm and underwent surgical intervention; the operator found a fistula connecting the aneurysm with PA. This finding was compatible with CAAAF.

Coronary artery fistulas are uncommon in patients who undergo coronary angiography. Fistulas from the RCA are slightly more common than those from the left coronary arteries. The reported incidence of bilateral coronary fistulas is 4-5%.¹⁰ It is rare to have RCA aneurysm with one fistula originating from it and the other one originating from the LAD.

In our case, although the patient had no objective evidence of myocardial ischemia, she presented anginal symptoms and dyspnea on exertion. The turbulent flow in the big aneurysm may alter the blood flow into distal vascular beds that may relate to the patient's clinical symptoms. A bulging shadow over right hilar region on chest radiograph was the initial image presentation. Subsequent studies including chest CT and MRI demonstrated a huge RCA aneurysm with compression of RV outflow tract. The decision for surgical intervention was made for the following reasons: symptomatic coronary aneurysm; the huge aneurysm may predispose to thrombus formation, resulting in myocardial ischemia or infarction; and the huge aneurysm continues to expand and may further compress RV outflow tract. The patient's symptoms completely resolved after surgical intervention.

Because of the relatively uncommon nature of these aneurysms, management of these patients has been based on anecdotal reports and experience, which included antiplatelet and anticoagulant medications. It is gener-

ally agreed that coronary artery bypass surgery should be performed in patients who have aneurysmal disease concomitant with significant coronary stenosis. Some authors recommended that ligation of the segment distal to the aneurysm can prevent distal embolization or rupture, but others had different opinions.^{11,12}

The treatment regarding a surgical approach in patients who have a coronary aneurysm without significant occlusive disease may be more controversial. Longitudinal studies are mandatory to determine the efficacy of medical therapy and the ideal interventional treatment.

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一例罕見之巨大冠狀動脈瘤合併瘻管

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接受心導管檢查的病患中約有 0.15% 到 4.9% 可發現有冠狀動脈瘤。而動脈粥狀硬化約佔 50% 成人冠狀動脈瘤之病因。我們報告一例 64 歲女性病患於右冠狀動脈合併有冠狀動脈瘤及瘻管連接此動脈瘤及肺動脈，並有另一瘻管連接左前降冠狀動脈近端及肺動脈，病患接受成功的冠狀動脈瘤切除及冠狀動脈瘻管結紮手術。此病患因右冠狀動脈巨大之動脈瘤並壓迫到右心室血流出口處臨床上表現為喘氣及類似心絞痛之症狀，使得本案例值得注意。病患的症狀也在手術後完全緩解。

關鍵詞：冠狀動脈瘤、冠狀動脈粥狀硬化、冠狀動脈瘻管。