

Single Left Coronary Artery With Microfistula Communicating With the Left Ventricle: A Case Report

Yoichi UECHI, MD

Koichi HIGA, MD

Abstract

A 79-year-old man with lung cancer undergoing chemotherapy and radiation complained of chest pain. Coronary angiography revealed that the right coronary artery arose from the distal branch of the left circumflex artery. In addition, a left coronary artery-to-left ventricular microfistula was recognized coincidentally. Multidetector row computed tomography revealed no other cardiac anomalies besides the single left coronary artery. No evidence of ischemia was noted during exercise stress myocardial scintigraphy. No vasospasm provocation study was performed, as it was considered to be potentially life-threatening for a patient with the single coronary system. In general, single coronary artery may occasionally represent a potentially fatal condition, so careful attention must be paid to its anatomical features.

J Cardiol 2007 Sep; 50(3):199–203

Key Words

■ Congenital heart disease

■ Coronary vessels

■ Angiography

INTRODUCTION

Single coronary artery, arising from the aortic trunk from a single coronary ostium and supplying the entire heart, is a rare congenital anomaly of the coronary circulation.^{1–5)} Coronary artery-to-left ventricular microfistula is also an uncommon form^{6–8)} of the various types of congenital coronary arterial fistulas reported.^{3–5)} Single coronary artery is occasionally associated with other congenital cardiac malformations.^{1,2,9)} Here we report a very rare case of single coronary artery coexisting with left coronary artery-to-left ventricular microfistula.

CASE REPORT

A 79-year-old man was referred to our hospital for the evaluation of chest pain. He was undergoing chemotherapy and radiation under a diagnosis of squamous cell carcinoma of the left lung. He complained of shortness of breath on exertion and had an approximately 2 months' history of suffering from recurrent episodes of rest angina at night, which was sometimes relieved by sublingual nitro-

glycerin. He had no coronary risk factors other than smoking habit.

Electrocardiography revealed supraventricular tachycardia or ventricular premature contractions, which were exacerbated by exertion. However, no significant ischemic changes could be documented at the time of chest pain. Physical examination revealed no abnormal heart murmurs. Chest radiography revealed the lung tumor in the left upper lung field, but no evidence of cardiac enlargement. Electrocardiography at rest showed biphasic or inverted T waves in leads II, III, aVF, and V₃–V₆, with frequent premature supraventricular contractions. Transthoracic two-dimensional echocardiography showed no regional wall motion abnormality or left ventricular hypertrophy. No significant valvular abnormalities were recognized.

Diagnostic coronary angiography was performed 4 days after admission. The left coronary artery was engaged and its ostium was located at its normal position in the left sinus of Valsalva. The origin and distribution of the main trunks on the left side, namely, the left anterior descending artery and the

牧港中央病院 内科: 〒901-2131 沖縄県浦添市牧港1199番地
Department of Internal Medicine, Makiminato Chuou Hospital, Okinawa

Address for correspondence: UECHI Y, MD, Department of Internal Medicine, Makiminato Chuou Hospital, Makiminato 1199, Urasoe, Okinawa 901–2131; E-mail: uechiy@r9.dion.ne.jp

Manuscript received March 26, 2007; revised May 14, 2007; accepted May 17, 2007

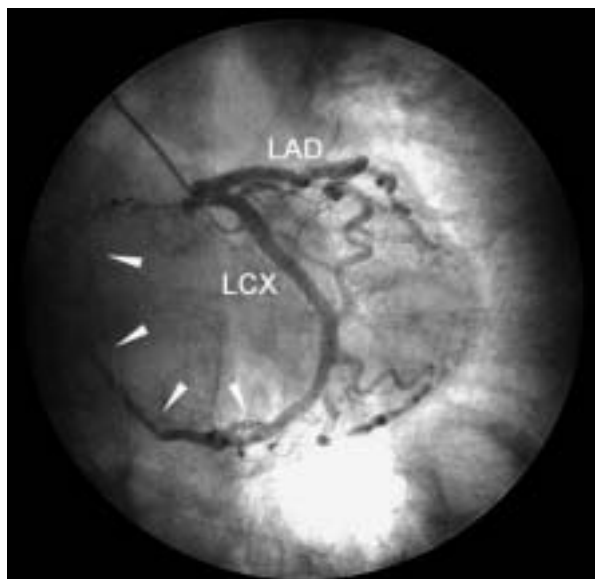


Fig. 1 Coronary angiogram in the anteroposterior projection with 30 degree caudal angulation

The right coronary artery arises from the distal branch of the left circumflex artery and continues retrogradely along the normal right coronary artery distribution (*arrowheads*).

LAD = left anterior descending artery; LCX = left circumflex artery.

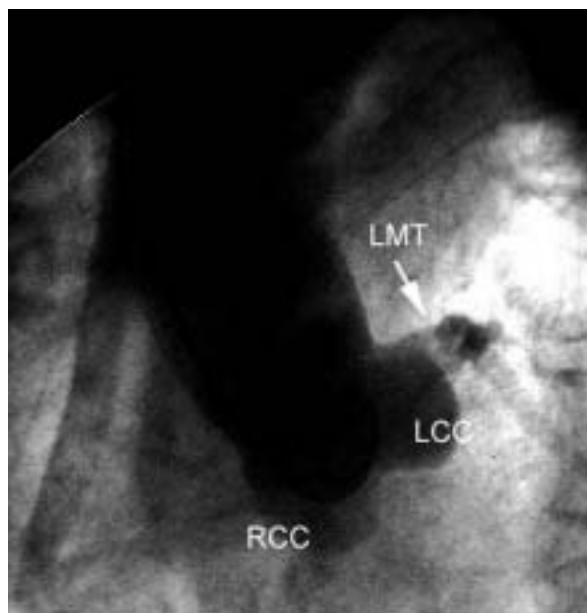


Fig. 2 Aortogram in the left anterior oblique view

Absence of the ostium of the right coronary artery is clearly confirmed.

LMT = left main coronary trunk; LCC = left coronary cusp; RCC = right coronary cusp.

left circumflex artery were normal, but the right coronary artery arose from the distal left circumflex artery, following the normal course of the right coronary artery distribution retrogradely. The distal portion was tapered, and terminated near the right sinus of Valsalva (**Fig. 1**). There was no evidence of stenotic lesions of the coronary arteries. Aortography confirmed the absence of the right coronary artery ostium (**Fig. 2**). Left ventriculography showed a normal-sized ventricle with an ejection fraction of 72%. Our routine catheterization study usually includes the acetylcholine provocation test to rule out vasospastic angina, but we decided against proceeding with this test because of the risk of life-threatening multi-vessel spasm in the patient with a single coronary system.

During the review of the angiograms, we noticed that after the injection of the contrast medium into the left coronary artery, the margin of the left ventricle also filled faintly at the end of diastole with the contrast medium arising diffusely from the left coronary artery, mainly the mid and distal portions of the left anterior descending artery (**Fig. 3**). We finally made a diagnosis of microfistula arising from the left coronary artery and draining into the

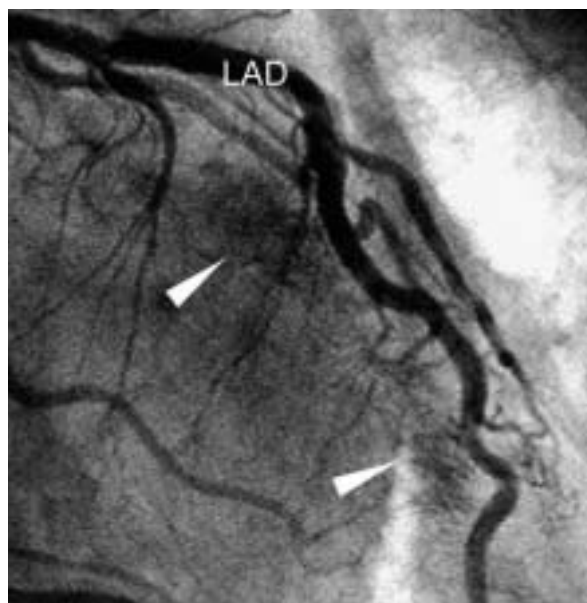


Fig. 3 Coronary angiogram in the right anterior oblique view

A small microfistula between the left anterior descending artery and the left ventricular cavity (*arrowheads*) is observed.

Abbreviation as in Fig. 1.

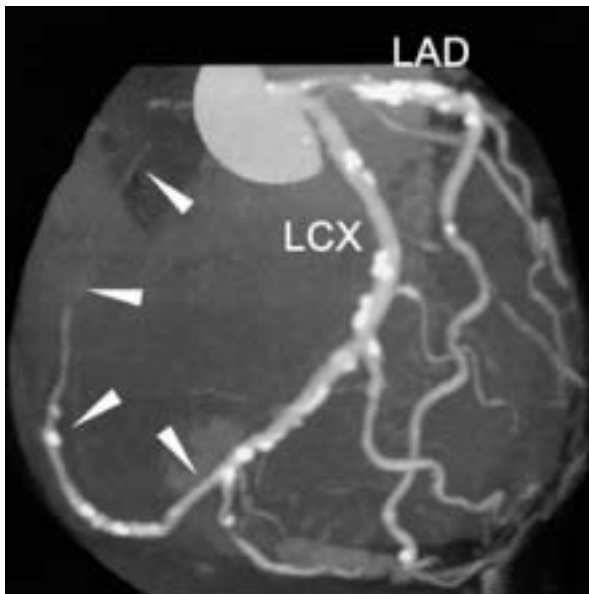


Fig. 4 Multidetector row computed tomography scan using maximum intensity projection

A single left coronary artery is visualized and the right coronary artery ostium is not visible (*arrowheads*). Calcified atherosclerotic lesions are also noted.

Abbreviations as in Figs. 1, 2.

left ventricular cavity.

Multidetector row computed tomography was performed to ascertain the diagnosis of the single coronary artery, which confirmed the presence of the single coronary artery (**Fig. 4**) and also the absence of any constriction of the coronary arteries by the great vessels (**Fig. 5**). However, the microfistula could not be visualized by this modality. Exercise stress myocardial scintigraphy using thallium-201 was performed to detect any evidence of myocardial ischemia. No symptomatic or significant electrocardiographic evidence of ischemia was noted during this test, and myocardial perfusion was not significantly impaired (**Fig. 6**).

The patient suffered no anginal attacks during the course of his stay at our hospital, and was discharged after a week of hospitalization with nitroglycerin medication, because vasospastic angina could not be ruled out at this time.

DISCUSSION

The incidence of single coronary artery with anomalous origin of the right coronary artery from the left circumflex artery is 0.02% to 0.04% in patients undergoing coronary angiography.¹⁻⁴ In our case, the right coronary artery originated from

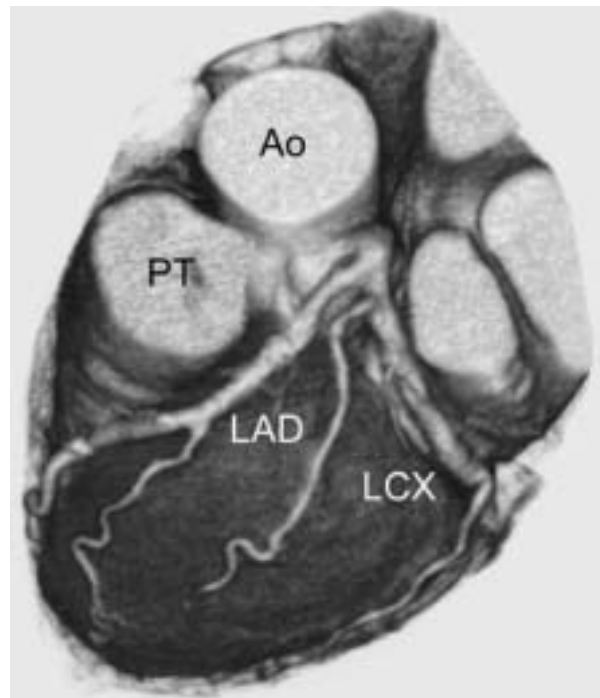


Fig. 5 Multidetector row computed tomography scan using volume rendering

The coronary arteries are not compressed between the aorta and the pulmonary trunk.

Ao = aorta; PT = pulmonary trunk. Other abbreviations as in Figs. 1, 2.

the distal left circumflex artery and crossed the crux to continue toward the right sinus of Valsalva. Several cases of this type of single coronary artery have been reported previously.^{1-4,9}

Single coronary artery is occasionally associated with other congenital cardiac anomalies, such as coronary arteriovenous fistula.^{1,2,9} In our case, a small coronary artery-to-left ventricular microfistula was accidentally identified coexisting with the single coronary artery. Coronary artery-to-left ventricular multiple microfistulas are defined as multiple microcommunications between one or more coronary arteries and the cavity of the left ventricle.⁸ Acquired coronary arterial fistula is rare but occurs after external thoracic trauma or as a consequence of surgery or myocardial biopsy.¹⁰ The present patient had no such past history, but the relationship between the coronary artery-to-left ventricular fistula and the lung cancer after radiotherapy was unclear.

Microfistulas between one or more coronary arteries and the left ventricular cavity have an incidence of 0.06%.⁸ Previously, a case of single right

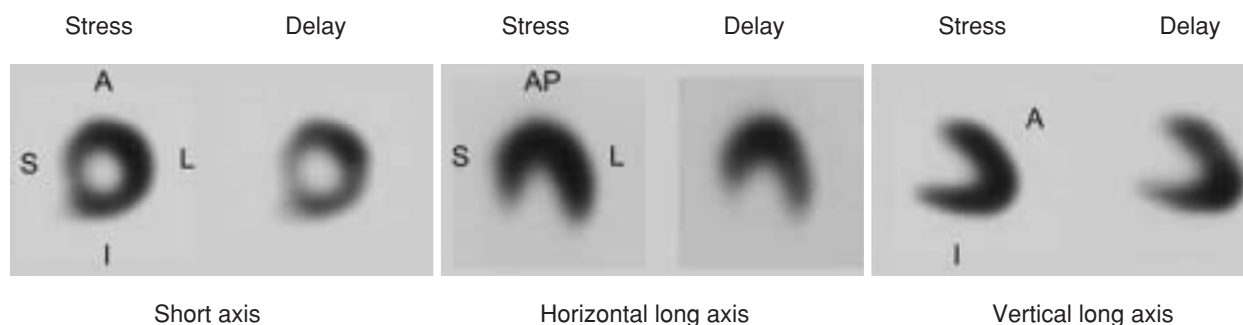


Fig. 6 Cardiac radionuclide single photon emission computed tomography scan using thallium-201

There is no significant exercise-induced impairment of myocardial perfusion.

S = septal; A = anterior; L = lateral; I = inferior; AP = apical.

coronary artery with left circumflex artery-to-left ventricular fistula was reported,¹¹⁾ but the present case of single left coronary artery with left coronary artery-to-left ventricular microfistula is unique. The coronary artery-to-left ventricular microfistula is usually small, in contrast to the more commonly encountered congenital fistulas between the coronary arteries and right heart structures (right atrium, right ventricle or pulmonary artery).¹⁰⁾ The small size of this type of coronary fistula is attributed to the similar diastolic pressures in the coronary capillary bed and the left ventricular cavity.¹²⁾ Although the pathophysiologic origin of fistulas draining into the left ventricle is obscure, morphological studies suggest that interference with developmental changes might produce an abnormally prominent thebesian system with the morphological appearance of a coronary microfistula.¹³⁾

Patients with single coronary artery are sometimes symptomatic, and some of these anomalies are associated with ischemic events and sudden death.^{1,3,14,15)} Several mechanisms of myocardial ischemia are possible in cases of single coronary artery, such as entrapment of an anatomically ectopic artery between the aorta and the pulmonary trunk, coronary vasospasm, and atherosclerotic changes in the coronary artery.^{1,3,14,15)} In our case, coronary angiography showed no significant organic stenoses and multidetector row computed tomography revealed no evidence of compression of the

coronary artery between the great vessels. Although we could not definitely exclude coronary spasm, our case appeared to be the benign type of single coronary artery.

The acetylcholine or ergonovine test is usually needed to evaluate coronary spasm. A patient with single coronary artery suffered vasospasm of the aberrant coronary artery induced by ergonovine.¹⁴⁾ At the time of the catheterization study, we decided against the test to provoke coronary arterial spasm because vasospasm of the single coronary system might result in the same situation of a life-threatening multi-vessel spasm. Therefore, we could not rule out vasospasm, but a plausible reason for the symptoms in our case was chronic insufficient blood flow from the abnormally distributed atherosclerotic vessels and resultant ischemic myocardium, exacerbated by the underlying coronary steal phenomenon caused by the microfistula.

While we could not find any evidence of myocardial ischemia, we considered that the symptoms in our patient could be of cardiac origin, and that the patient should remain under careful observation because of the risk of future ischemic events with the progression of coronary atherosclerosis or spontaneous vasospasm in his single coronary system.

From the clinical point of view, awareness of such anomalous origins of the coronary arteries is important to establish the precise diagnosis and make appropriate therapeutic decisions based on coronary angiography.

要 約

左冠動脈左室瘻を合併した左単冠動脈の1例

上地 洋一 比嘉 耕一

右冠動脈が左回旋枝から起始している左単冠動脈に、左冠動脈左室瘻を合併したまれな症例を経験したので報告する。症例は79歳の男性で、肺癌に対し化学療法と放射線療法を受けていた。胸痛の訴えがあったため、冠動脈疾患の精査を目的に心臓カテーテル検査を行った。冠動脈造影検査の結果、左冠動脈の起始部およびその分枝は解剖学的に正常であったが、右冠動脈が左回旋枝の遠位部より起始していることが明らかとなった。さらに、左冠動脈左室瘻の合併も認められた。マルチスライスコンピューター断層撮影上、左単冠動脈以外の心奇形は認められなかった。運動負荷心筋シンチグラフィにおいて、心筋虚血は認められなかった。冠動脈の攣縮誘発試験は、単冠動脈の患者に対しては致命的な不整脈を惹起する危険性が高いと思われ行わなかった。一般的に、単冠動脈の中には致命的な症例もあることから、その解剖学的な特徴には注意を払うべきである。

J Cardiol 2007 Sep; 50(3): 199–203

References

- 1) Chaitman BR, Lespérance J, Saltiel J, Bourassa MG: Clinical, angiographic, and hemodynamic findings in patients with anomalous origin of the coronary arteries. *Circulation* 1976; **53**: 122–131
- 2) Lipton MJ, Barry WH, Obrez I, Silverman JF, Wexler L: Isolated single coronary artery: Diagnosis, angiographic classification, and clinical significance. *Radiology* 1979; **130**: 39–47
- 3) Wilkins CE, Betancourt B, Mathur VS, Massumi A, De Castro CM, Garcia E, Hall RJ: Coronary artery anomalies: A review of more than 10,000 patients from the Clayton Cardiovascular Laboratories. *Tex Heart Inst J* 1988; **15**: 166–173
- 4) Yamanaka O, Hobbs RE: Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn* 1990; **21**: 28–40
- 5) Aydinlar A, Çiçek D, Sentürk T, Gemici K, Serdar OA, Kazazoglu AR, Kumbay E, Cordan J: Primary congenital anomalies of the coronary arteries: A coronary arteriographic study in Western Turkey. *Int Heart J* 2005; **46**: 97–103
- 6) Nawa S, Miyachi Y, Toshino N, Shiba T, Hayashi K, Tamesue K, Yamamoto H, Shimizu N: Three major coronary artery-to-left ventricular shunts: Report of three cases and review of literature. *Cardiovasc Intervent Radiol* 1997; **20**: 300–304
- 7) Stierle U, Giannitsis E, Sheikhzadeh A, Potratz J: Myocardial ischemia in generalized coronary artery-left ventricular microfistulae. *Int J Cardiol* 1998; **63**: 47–52
- 8) Said SA, van der Werf T: Dutch survey of congenital coronary artery fistulas in adults: Coronary artery-left ventricular multiple micro-fistulas Multi-center observational survey in the Netherlands. *Int J Cardiol* 2006; **110**: 33–39
- 9) Ogden JA, Goodyer AV: Patterns of distribution of the single coronary artery. *Yale J Biol Med* 1970; **43**: 11–21
- 10) Gillebert C, Van Hoof R, Van de Werf F, Piessens J, De Geest H: Coronary artery fistulas in an adult population. *Eur Heart J* 1986; **7**: 437–443
- 11) Brito FS Jr, Vianna CB, Caixeta AM, Rati MA, Perin MA, Ramires JA, Martinez Filho EE: Single coronary arteries: Two cases with distinct and previously undescribed angiographic patterns. *J Invasive Cardiol* 1999; **11**: 430–434
- 12) Cheng TO: Left coronary artery-to-left ventricular fistula: Demonstration of coronary steal phenomenon. *Am Heart J* 1982; **104**: 870–872
- 13) Black IW, Loo CK, Allan RM: Multiple coronary artery-left ventricular fistula: Clinical, angiographic, and pathophysiologic findings. *Cathet Cardiovasc Diagn* 1991; **23**: 133–135
- 14) Yamamoto K, Koizumi Y, Tajimi T, Inou T, Mitsutake A, Orita Y, Takeshita A, Nakamura M: Coronary arterial spasm in single coronary artery. *Circulation* 1981; **64**: 1287–1290
- 15) Taylor AJ, Rogan KM, Virmani R: Sudden cardiac death associated with isolated congenital coronary artery anomalies. *J Am Coll Cardiol* 1992; **20**: 640–647