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Pitfalls of Intervention Therapy in a Patient With Anomalous Origin of the Right Coronary Artery From the Left Sinus of Valsalva Associated With Organic Stenosis

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Abstract

A 64-year-old man visited our hospital with a complaint of exertional chest discomfort. Exercise electrocardiography revealed ST segment depression in the V_4–V_6 leads, and exercise thallium myocardial scintigraphy demonstrated myocardial ischemia in the area of the right coronary artery, suggesting effort angina. Diagnostic coronary angiography revealed an anomalous origin of the right coronary artery from the left sinus of Valsalva and 90% organic stenosis at the proximal portion. We performed percutaneous transluminal coronary angioplasty (PTCA), but repeat PTCA was required 3 months later because of restenosis. Follow-up angiography 1 year later showed regression of the stenotic lesion to less than 50% diameter compared with the data obtained 3 months after the second PTCA. However, exercise ^{99m}technetium-tetrofosmin myocardial scintigraphy disclosed obvious myocardial ischemia in the inferior region.

These results suggested that the myocardial ischemia in this particular patient was caused not only by the organic stenosis but also by the anatomic anomaly which might reduce coronary blood flow during exercise. Such patients should be followed up cautiously with much more sophisticated methodology.

Key Words

Angioplasty (percutaneous transluminal coronary), Radionuclide imaging (99m-Tc-tetrofosmin myocardial scintigraphy), Coronary circulation, Revascularization, Exercise tests, Congenital heart disease (anomalous origin of the right coronary artery)

INTRODUCTION

Anomalous origin of the coronary artery is a rare congenital anomaly with an incidence of about 0.3 to 1.2%, and anomalous origin of the right coronary artery from the left sinus of Valsalva has been found in 6–27% of patients with this anomaly[1,2]. This anomaly was thought to be a clinically benign abnormality, but it was reported to cause angina pectoris or myocardial infarction in the absence of any distinct atherosclerotic lesion, and also to cause faintness, ventricular fibrillation and even sudden death[3–5].

We encountered a patient with this anomaly asso-

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Selected abbreviations and acronyms

PTCA = percutaneous transluminal coronary angioplasty
Tc = technetium
Tl = thallium

ciated with atherosclerotic stenosis, who underwent elective percutaneous transluminal coronary angioplasty (PTCA) for myocardial ischemia. However, follow-up examination showed that the anomalous right coronary artery was related to the mechanism responsible for the development of myocardial ischemia and we were obliged to change the therapeutic strategy. Therefore, we wish to propose the most appropriate therapeutic strategy for such cases complicated by organic stenosis.

CASE REPORT

A 64-year-old man had developed persistent exertional chest discomfort for several minutes for 2 weeks before he visited our hospital in June 1994. Exercise electrocardiography revealed ST segment depression in the V_4–V_6 leads and exercise myocardial scintigraphy demonstrated transient myocardial ischemia in the region of the right coronary artery. As these results were highly suggestive of effort angina, we performed diagnostic coronary angiography, which disclosed the normal left coronary artery and anomalous origin of the right coronary artery from the left sinus of Valsalva. Furthermore, the latter had 90% organic stenosis at the proximal portion. Therefore, we thought that the myocardial ischemia was caused by this atherosclerotic lesion. We therefore performed PTCA with satisfactory results.

Follow-up coronary angiography 3 months later showed 90% restenosis at the same portion, so PTCA was repeated. Further follow-up angiography revealed mild progression (50–75% narrowing) of the stenosis in comparison with that immediately after the second PTCA (**Fig. 1**). However, he had no apparent symptoms and exercise thallium-201 (Tl) myocardial scintigraphy showed a slight decrease of Tl uptake in the inferior region (**Fig. 2**). We decided to observe this patient conservatively.

About 1 year after the second PTCA, the patient began to complain of exertional chest discomfort again and exercise electrocardiography showed ST segment depression in the V_4–V_6 leads, so we re-

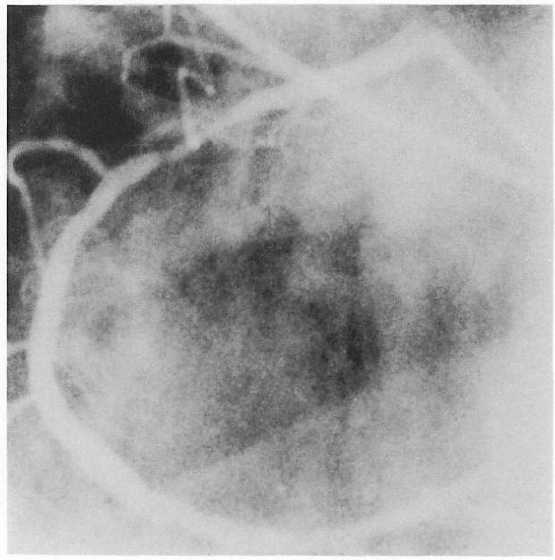

Fig. 1 Coronary angiogram 3 months after 2nd PTCA showing organic stenosis causing 50–75% narrowing at the proximal portion of the right coronary artery

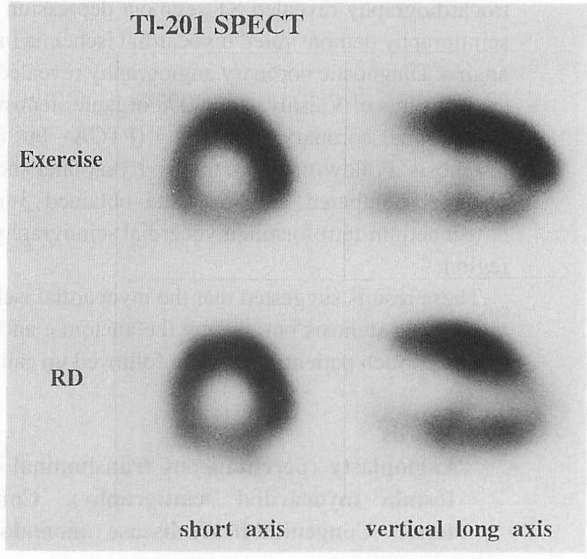

Fig. 2 Exercise thallium myocardial scintigrams 3 months after 2nd PTCA showing slight decrease of Tl uptake in the inferior region

examined the suspected stenosis. Contrary to our prediction, the stenosis had regressed to 50% in diameter (**Fig. 3**). Therefore, in addition to dipyridamole-stress Tl scintigraphy, exercise technetium-99m (^{99m}Tc)-tetrofosmin myocardial scintigraphy, which has higher energy than Tl, was performed to confirm whether the myocardial ischemia was present or not. Dipyridamole-stress Tl scintigraphy showed a slight decrease of Tl uptake in the inferior

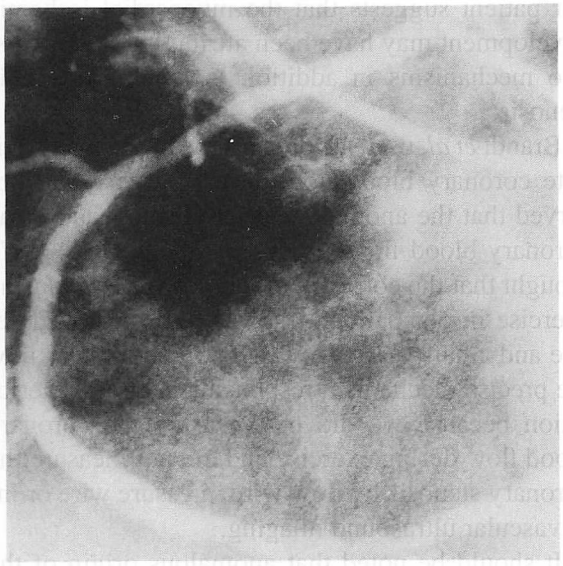

Fig. 3 Angiogram 1 year after 2nd PTCA showing the stenosis had regressed to 50% in diameter

region but no myocardial ischemia, whereas exercise ^{99m}Tc-tetrofosmin myocardial scintigraphy revealed obvious myocardial ischemia in the region of the right coronary artery (**Fig. 4**). Fast computed tomography of the patient revealed that the right coronary artery originated from the left sinus of Valsalva at an acute angle and coursed between the aorta and the pulmonary artery (**Fig. 5**).

Thus, we suspected the anomalous origin of this artery was related to the development of myocardial ischemia during exercise, and decided to perform bypass surgery to the right coronary artery.

DISCUSSION

It was reported that there is no difference in the ability to diagnose myocardial ischemia between dipyridamole stress and exercise Tl scintigraphy[4]. On the other hand, it has been thought that the use of exercise Tl scintigraphy may lead to overdiagnosis due to poor Tl uptake in the inferior region. ^{99m}Tc-tetrofosmin has higher radioactivity than Tl and is claimed to have similar sensitivity compared with that of exercise Tl scintigraphy using the reinjection method. Therefore, exercise ^{99m}Tc-tetrofosmin myocardial scintigraphy would be expected to reduce the possibility of overdiagnosis related to inferior myocardial ischemia[5,6].

In our patient, obvious myocardial ischemia was revealed by exercise ^{99m}Tc-tetrofosmin myocardial scintigraphy in spite of the regression of the atherosclerotic stenosis 1 year after the second PTCA. On the contrary, dipyridamole-stress Tl scintigraphy performed during the same period demonstrated slight reduction of Tl uptake but no abnormalities indicating myocardial ischemia in the inferior region. These results obtained by the two different myocardial scintigraphic methods showed that the myocardial ischemia in this case was not only caused by atherosclerotic stenosis, but also by functional ischemia due to exercise stress resulting from the anatomical anomaly.

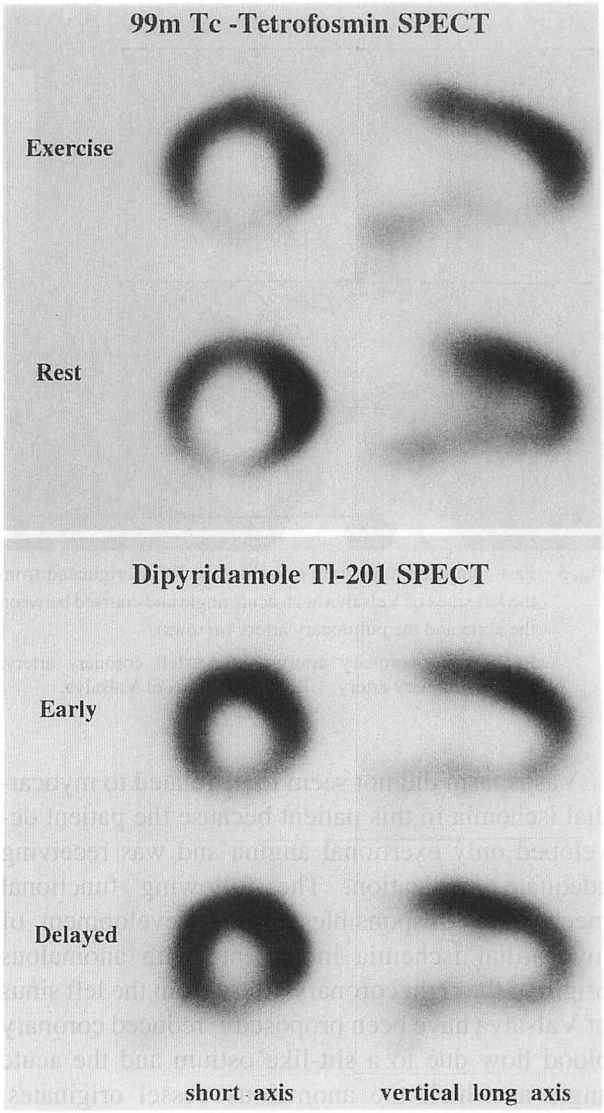

Fig. 4 Scintigrams 1 year after 2nd PTCA
Upper: Exercise ^{99m}Tc-tetrofosmin myocardial scintigram revealing obvious myocardial ischemia in the region of the right coronary artery.
Lower: Dipyridamole-stress Tl scintigram showing a slight decrease of Tl uptake in the inferior region but no myocardial ischemia.

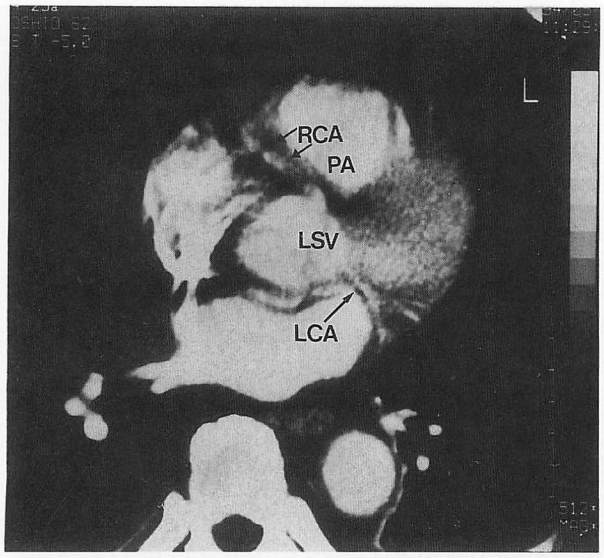

Fig. 5 Fast computed tomogram revealing the RCA originated from the left sinus of Valsalva at an acute angle and coursed between the aorta and the pulmonary artery (*arrows*)

RCA＝right coronary artery; LCA＝left coronary artery; PA＝pulmonary artery; LSV＝left sinus of Valsalva.

Vasospasm did not seem to be related to myocardial ischemia in this patient because the patient developed only exertional angina and was receiving adequate medication. The following functional mechanisms responsible for the development of myocardial ischemia in patients with anomalous origin of the right coronary artery from the left sinus of Valsalva have been proposed : reduced coronary blood flow due to a slit-like ostium and the acute angle at which the anomalous vessel originates, compression between the aorta and pulmonary trunk[7–9], and kinking due to the long run of the vessel[10]. All these speculative mechanisms were based on exercise stress. Fast computed tomography of our patient suggests that the myocardial ischemia development may have been attributable to the first two mechanisms in addition to the mild organic stenosis.

Brandt *et al.* used a Doppler flow probe to investigate coronary blood flow during surgery and observed that the anomalous origin itself reduced the coronary blood flow reserve by about 50%[11]. We thought that the coronary blood flow reserve during exercise in our patient gradually deteriorated over one and a half years. However, we could not know the precise mechanism responsible for this deterioration because we did not measure the coronary blood flow during exercise and did not measure the coronary stenosis by flow wire, pressure wire or intravascular ultrasound imaging.

It should be noted that anomalous origin of the right coronary artery from the left sinus of Valsalva is not a benign congenital anomaly as has been generally accepted, so patients with this anomaly should be assessed carefully using exercise myocardial scintigraphy. Even if the organic stenosis is mild (American Heart Association class : 50% or less) on angiograms, such patients should be followed up carefully, and once any reduction in the coronary blood flow reserve is found, after making a clear evaluation of the organic stenosis by not only coronary angiography, but also flow wire, pressure wire or intravascular ultrasound imaging, a complete operative revascularization should be considered. Furthermore, even after intervention therapy such as PTCA is performed to alleviate complicated organic stenosis, careful attention should be paid to the residual myocardial ischemia caused by this anomaly to avoid an inadequate therapeutic strategy.

要　約

有意狭窄を伴う右冠動脈左 Valsalva 洞起始症の 1 例：インターベンションの適応判定における落し穴

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症例は 64 歳，男性．胸部圧迫感を訴え来院．負荷心電図で V_4–V_6 の ST 低下，運動負荷 Tl 心筋シンチグラフィーで右冠動脈領域の虚血を認め，労作性狭心症を疑い，冠動脈造影を施行した．その結果，右冠動脈左 Valsalva 洞起始症と同冠動脈近位部の 90% 器質的狭窄を認め，PTCA を施行した．しかし，再狭窄のため，3 ヵ月後に再 PTCA を施行することとなった．再 PTCA 施行 1 年後の冠動脈造影では再PTCA 3 ヵ月後に比し，病変部は 50% 狭窄に改善し，dipyridamole

負荷心筋シンチグラフィーでは下壁領域に若干の取り込み低下を認めるのみであったが，運動負荷 ^{99m}Tc-tetrofosmin 心筋シンチグラフィーでは下壁領域に明らかな心筋虚血の増悪が証明された．

以上の結果から，本例での心筋虚血は，冠動脈の器質的有意狭窄のみならず，右冠動脈左 Valsalva 洞起始症という解剖学的奇形に運動負荷が加わり，惹起されたと考えられる．本奇形においては，冠動脈造影上，器質的狭窄度が50％程度しかなくとも，運動負荷時冠血流予備能の評価という点で慎重な運動負荷シンチグラフィーでの経過観察が必要であり，また必要に応じ，より詳細な検討を加える必要があろう．

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References

1) Taylor AJ, Rogan KM, Virmani R : Sudden cardiac death associated with isolated congenital coronary artery anomalies. J Am Coll Cardiol 1992; **20** : 640–647
2) Kimbiris D, Iskandrian AS, Segal BL, Bemis CE : Anomalous aortic origin of coronary arteries. Circulation 1978; **58** : 606–615
3) Benge W, Martins JB, Funk DC : Morbidity associated with anomalous origin of the right coronary artery from the left sinus of Valsalva. Am Heart J 1980; **99** : 96–100
4) Leppo JA : Dipyridamole-thallium imaging : The lazy man's stress test. J Nucl Med 1989; **30** : 281–287
5) Zaret BL, Rigo P, Wackers FJT, Hendel RC, Braat SH, Iskandrian AS, Sridhara BS, Jain D, Itti R, Serafini AN, Goris ML, Lahiri A : The Tetrofosmin International Trial Study Group : Myocardial perfusion imaging with ^{99m}Tc tetrofosmin : Comparison to ^{201}Tl imaging and coronary angiography in a phase III multicenter trial. Circulation 1995; **91** : 313–319
6) Nishimura T, Nobuyoshi M : Clinical application of ^{99m}Tc tetrofosmin myocardial SPECT : A multicenter trial. Jpn J Nucl Med 1995; **32** : 1007–1021 (in Japanese)
7) Cheitlin MD, De Castro CM, McAllister HA : Sudden death as a complication of anomalous left coronary origin from the anterior sinus of Valsalva: A not-so-minor congenital anomaly. Circulation 1974; **50** : 780–787
8) Pachinger OM, VandenHoven P, Judkins MP : Single coronary artery : A cause of angina pectoris. Eur J Cardiol 1974; **2** : 161–165
9) Virmani R, Chun PK, Goldstein RE, Robinowitz M, McAllister HA : Acute takeoffs of the coronary arteries along the aortic wall and congenital coronary ostial valve-like ridges : Association with sudden death. J Am Coll Cardiol 1984; **3** : 766–771
10) Jokl E, McClellan JT, Ross GD : Congenital anomaly of left coronary artery in young athlete. JAMA 1962; **182** : 572–573
11) Brandt B III, Martins JB, Marcus ML : Anomalous origin of the right coronary artery from the left sinus of Valsalva. N Engl J Med 1983; **31** : 596–598