ELSEVIER

International Journal of Cardiology 126 (2008) e13–e16

International Journal of Cardiology

www.elsevier.com/locate/ijcard

Letter to the Editor

Catheter-based management for the congenital coronary arteriovenous fistula indicated by the stress ^{99m}Tc-MIBI SPECT

Meng-Luen Lee[a,*], Jui-Wen Peng[b], Guang-Uei Hung[c]

[a] *Department of Pediatrics, Division of Pediatric Cardiology, Changhua Christian Hospital, No. 135, Nanhsiao St., Changhua 50050, Taiwan*
[b] *Department of Medicine, Cardiovascular Laboratory, Cardiac Catheterization, Changhua Christian Hospital, Changhua 50050, Taiwan*
[c] *Department of Nuclear Medicine, Changhua Christian Hospital, Changhua 50050, Taiwan*

Received 18 November 2006; accepted 31 December 2006
Available online 17 April 2007

Abstract

A 10-year-old boy, who had suffered Kawasaki disease 7 years ago, presented chest pain, pale face, and cold sweating in the stress ^{99m}technetium-methoxyisobutylisonitrile (^{99m}Tc-MIBI) single-photon emission computed tomography (SPECT), which showed reversible perfusion abnormality indicative of myocardial ischemia involving the right coronary artery. Angiography identified congenital coronary arteriovenous fistula (CAVF) from the right coronary artery to the main pulmonary artery, to which the patient's symptoms and signs of myocardial ischemia, by means of coronary steal phenomenon that entails reversible perfusion abnormality in the stress ^{99m}Tc-MIBI SPECT, could be ascribed. The penny shall finally be dropped without sudden cardiac event after coaxial transarterial coil occlusion on this boy with the congenital CAVF that is indicated significantly by the stress ^{99m}Tc-MIBI SPECT.

Keywords: Kawasaki disease; Chest pain; ^{99m}Technetium-methoxyisobutylisonitrile; Single-photon emission computed tomography; Coronary arteriovenous fistula; Coil occlusion

Dipyridamole-thallium myocardial single-photon emission computed tomography (SPECT) has been recommended for risk stratification in the long-term follow-up of patients with Kawasaki disease (KD) [1], in that SPECT can be non-invasively and safely performed in young children who are unable to exercise adequately [1,2]. We performed stress ^{99m}technetium-methoxyisobutylisonitrile (^{99m}Tc-MIBI) myocardial SPECT to assess coronary arterial sequelae on a 10-year-old boy, who had suffered KD 7 years ago, presenting chest pain, cold sweating, and pale face after 100-meter dashing and long-distance jogging in recent 3 months. He complained chest pain and presented pale face and cold sweating in the stress ^{99m}Tc-MIBI SPECT, which showed reversible perfusion abnormality indicative of myocardial ischemia in the territories of the right coronary artery. Angiography identified congenital coronary arteriovenous fistula (CAVF) from the right coronary artery to the main pulmonary artery, to which patient's symptoms and signs of myocardial ischemia by coronary steal phenomenon could be ascribed. The penny shall finally be dropped without sudden cardiac event in this boy after coaxial transarterial coil occlusion for the insidious congenital CAVF. Catheter-based management for the congenital CAVF indicated significantly by the stress ^{99m}Tc-MIBI SPECT has never been reported in the English literature.

A 10-year-old boy had suffered from KD at the age of 3 years old. At that time he received one standard dose of intravenous gamma-globulin (2 gm/kg) and high-dose oral aspirin (80 mg/kg/day). Since discharge from the initial hospitalization, he was free of symptoms and signs in the 7-year follow-up for KD. However, he complained fatigability and chest pain, and presented cold sweating and pale face after 100-meter dashing and long-distance jogging, in recent 3 months. Stress/rest ^{99m}Tc-MIBI SPECT with a bicycle ergometer, which was performed to assess coronary arterial sequela of KD, showed reversible perfusion

* Corresponding author. Tel.: +886 4 7238595x1035; fax: +886 4 7238847.
E-mail address: ferdielee@yahoo.com (M.-L. Lee).

0167-5273/$ - see front matter © 2007 Elsevier Ireland Ltd. All rights reserved.
doi:10.1016/j.ijcard.2006.12.072

abnormality in the basal inferoseptal segment, the basal inferior segment, the mid inferior segment, and the apical inferior segment that significantly indicate myocardial ischemia involving the right coronary artery (Fig. 1). Follow-up echocardiography revealed turbulent flow in an abnormal vessel draining to the main pulmonary artery. However, we could not trace it back to its origin. The ascending aortography and the selective right coronary angiography finally tracked down the culprit to be a congenital CAVF draining the right coronary artery to the main pulmonary artery (Fig. 2A–D). The pulmonary-to-systemic blood flow ratio is 1.16. This CAVF was subsequently occluded (Fig. 2E, F) by 3 sets of the Complex Helical Fibered Platinum Coil-18 (TARGET, Medi-tech, Watertown, MA, USA), which were delivered retrogradely by a Vortx Coil Pusher-16 (TARGET, Medi-tech, Watertown, MA, USA) via a coaxial system [3], which is composed of a smaller Tracker-18MX Infusion Catheter (TARGET, Medi-tech, Watertown, MA, USA) housed coaxially into a larger 5-Fr Right Coronary Judkins Catheter. There was no more fatigability, chest pain, cold sweating, and pale face in the follow-up stress/rest ^{99m}Tc-MIBI SPECT, which documents regression of myocardial ischemia in the aforementioned segments of the right coronary artery. He was free of symptoms and signs of myocardial ischemia at the 12-month follow-up in the outpatient clinic of the Division of Pediatric Cardiology.

1. Discussion

Congenital CAVF is a rare congenital anomaly that may cause severe morbidity and mortality [4]. The natural history and the clinical course of which vary significantly among affected patients. Generally speaking, the symptoms develop depending on the amount of the shunt or the presence of coronary steal phenomenon of the fistulas, which tend to present in infants below 2 years of age with congestive heart failure, to develop in young adults with angina, exertional dyspnea, myocardial ischemia, and infarction, and to manifest in adults >40 years old with congestive heart failure, atherosclerosis, and cardiac arrhythmias [4]. Though some patients may have symptoms shortly after birth [5], however most of them are asymptomatic [6].

Congenital CAVF drains most commonly to the right cardiac chambers or vessels (right ventricle, right atrium, pulmonary artery, and coronary sinus in decreasing frequency) with lower blood pressures. The most important clue of diagnosis is the location and characteristic of cardiac murmur, which could usually be appreciated along the right sternal border as continuous murmur on auscultation [3]. However, this leading clue could be absent with a small fistulous shunt. Although congenital CAVF can be diagnosed by Doppler echocardiography with color flow mapping [7], but a harbinger of which in the present case are symptoms and signs of myocardial ischemia provoked by exercise and by the stress ^{99m}Tc-MIBI SPECT, rather than a physical sign of continuous murmur by auscultation.

Congenital CAVF with a pulmonary-to-systemic blood flow ratio (Qp/Qs) >2.0 and hemodynamic change and symptoms of heart failure are recommended for surgery [8]. However, Sato and Koishizawa reported one case with congenital CAVF, in spite of having a low Qp/Qs ratio of 1.08, developing symptoms of myocardial ischemia after the

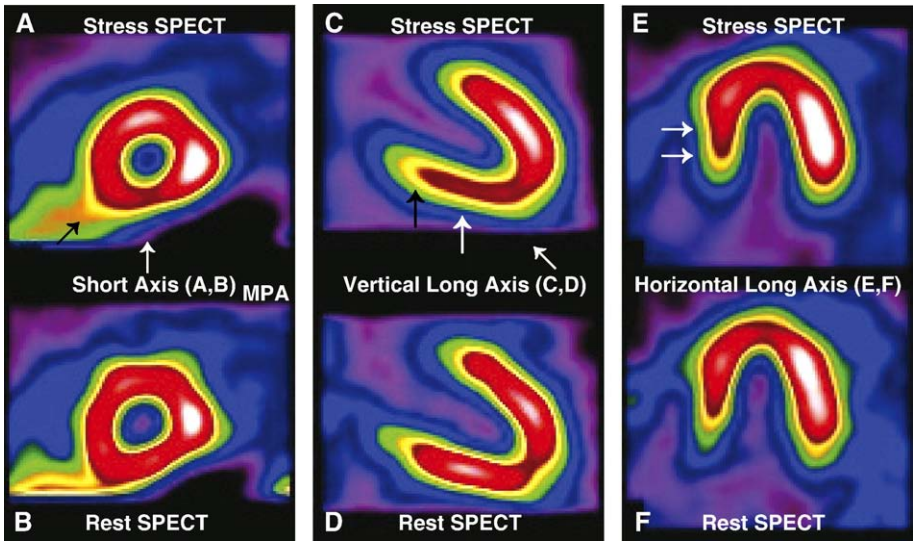

Fig. 1. Stress/rest ^{99m}Tc-MIBI SPECT through the short axis (A, B), the vertical long axis (C, D), and the horizontal long axis (E, F) of heart. Stress ^{99m}Tc-MIBI SPECT showed perfusion abnormalities in the basal inferior segment (vertical arrow, A), the basal inferoseptal segment (oblique arrow, A), the apical inferior segment (oblique arrow, C), the mid inferior segment (vertical white arrow, C), the basal inferior segment (vertical black arrow, C), and the basal inferoseptal segment (horizontal arrows, E), which are assigned to the territory of the right coronary artery. All these perfusion abnormalities are illustrated to be reversible at rest ^{99m}Tc-MIBI SPECT (B, D, and F). The aforementioned segments 3 (basal inferoseptal segment), 4 (basal inferior segment), 10 (mid inferior segment), and 15 (apical inferior segment) are accordingly assigned to the right coronary artery [12].