Magnetic Resonance Imaging of Bland-White-Garland Syndrome

— A Case of Anomalous Origin of the Left Coronary Artery From the Pulmonary Trunk in a 22-Year-Old Woman —

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Selective right coronary angiography in a 22-year-old woman demonstrated an enormous right coronary artery, with the contrast medium opacifying to the left coronary artery through a well-developed collateral circulation and draining into the pulmonary artery. The diagnosis of anomalous origin of the left coronary artery from the pulmonary trunk [also known as Bland-White-Garland (BWG) syndrome] was thus established. Magnetic resonance imaging (MRI) showed a dilated right coronary artery arising from the aorta, the left coronary artery from the pulmonary trunk but not from the aorta, and well-developed channels in the interventricular septum. MRI could be a useful tool for diagnosing anomalous origin of the left coronary artery from the pulmonary trunk. (*Jpn Circ J* 1998; 62: 219–221)

Key Words: Bland-White-Garland syndrome; Congenital heart disease; Magnetic resonance imaging

A left coronary artery originating from the pulmonary trunk, often referred to as Bland-White-Garland (BWG) syndrome, is a rare congenital heart disorder. Most patients with this anomaly die in infancy! However, 5–10% survive an early myocardial infarction, with subsequent development of coronary collateral vessels from the right coronary artery to the left coronary artery? Many such patients are asymptomatic, and the establishment of this anomaly is often difficult.

In this report, we describe an adult patient in whom magnetic resonance imaging (MRI) clearly demonstrated the anomalous origin of the left coronary artery.

Case Report

A 22-year-old woman was referred to us because of exertional palpitation. On physical examination, a 1/6 systolic murmur along the left lower sternal border was the only finding. An electrocardiogram showed a small q wave in leads I and aVL. A treadmill exercise test showed horizontal ST-segment depression in leads II, III, aVF, and V₃–V₆ but the patient experienced no chest pain. Two-dimensional and color Doppler echocardiography identified a normal-sized left ventricle with hypokinesis of the anterior wall, and Doppler flow was shown in diastole from the lower to upper portion of the interventricular septum. Exercise [^{99m}Tc]-tetrofosmin scintigraphy showed transient and severe hypoperfusion

in the anterior wall. Selective right coronary angiography demonstrated an enormous right coronary artery, and the left coronary artery was revealed by the contrast flow through the collateral circulation, showing the anomalous origin from the pulmonary artery (Fig 1, top). The aortogram showed only the right coronary artery arising from the aorta, and no left coronary artery (Fig 1, bottom). The pressure values in the cardiac chambers, pulmonary artery, and aorta were all within normal ranges, and the estimated left-to-right shunt, pulmo-systemic flow ratio (Q_p/Q_s) was 1.81.

MRI performed with an ECG-gated multislice spin-echo sequence showed an anatomically normal relationship of chambers and great vessels, a dilated right coronary artery arising from the aorta (Fig 2A, C and D), well-developed vessels in the interventricular septum (Fig 2D), and the left coronary artery from the pulmonary trunk but not from the aorta (Fig 2B).

The diagnosis of anomalous origin of the left coronary artery from the pulmonary trunk with old myocardial infarction of a small area of anterior wall and myocardial ischemia was established. The cause of exertional palpitation was undetermined. The patient is being followed up with careful observation in our outpatient department until surgical repair can be performed.

Discussion

Most patients with anomalous origin of the left coronary artery from the pulmonary trunk die in infancy. In a clinical spectrum, pathological, and pathophysiological study, Wesselhoeft et al¹ suggested that several mechanisms, including the development of an interarterial collateral circulation, altered hemodynamics that would encourage flow into the vascular bed, and reduction in

(Received May 30, 1997; revised manuscript received August 11, 1997; accepted August 26, 1997)

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the area of the myocardium supplied by the anomalous left coronary artery were important for survival beyond infancy. Such surviving patients have developed collateral vessels and a dilated right coronary artery arising from the aorta. To our knowledge, only 2 cases of left coronary

Fig 1. (A) Right anterior oblique right coronary arteriogram. (B) Left anterior oblique aortogram. (A) demonstrates the enormous right coronary artery. The left coronary artery is opacified by retrograde contrast flow from the right coronary artery through the well-developed collateral, and the contrast drains into the pulmonary artery (white arrow). (B) shows the right coronary artery arising from the aorta but no left coronary artery.

artery originating in the pulmonary trunk have been previously studied by MRI.^{3,4} The previously reported patients and our patient were all adults with developed collateral vessels. In our patient, MRI demonstrated dilated right coronary artery from the aorta, the anomalous origin of the left coronary artery from the pulmonary trunk, and developed vessels in the interventricular septum. The first 2 findings were similar to those of the previous reports.^{3,4} Vascular channels connecting the right and left coronary arteries were reported on the surface of the heart.⁴ In our patient, the vascular channels depicted in the interventricular septum on MRI coincided with the findings of Doppler flow in the interventricular septum on 2-dimensional and color Doppler echocardiography.

Several types of important information can be derived from MRI, an entirely non-invasive imaging technique. Although spin-echo MRI gives detailed anatomic information and depicts rapidly moving blood as a signal void, the ability of MRI to delineate the coronary artery structure may be unsatisfactory. On occasion, the anatomy of the aortic root, the coronary ostia, and the major proximal portions of the coronary arteries have been documented in particular conditions with significant coronary dilatation, such as fistulae and aneurysms.⁵ However, recent progress from conventional spin-echo imaging to high-speed MRI methods, including the fast gradient-echo technique with time of flight or phase-contrast method, the fast spin-echo technique, and the echo planar imaging technique, as well as the further development of several MRI techniques has led to the clinical use of these techniques in evaluating coronary morphology and flow.^{6,7} Manning et al⁸ reported the possibility of imaging of the proximal, mid, and distal epicardial coronary arteries by breath-hold magnetic resonance coronary angiographic techniques. Recently, the imaging of other types of anomalous coronary arteries using these spin-echo or developed magnetic resonance coronary angiographic techniques has been reported.⁹⁻¹¹

Therefore, in adult patients with anomalous origin of the left coronary artery from the pulmonary artery with developed interarterial collateral circulation, MRI is a useful tool for diagnosis. Moreover, with further improvements in magnetic resonance hardware and software, MRI appears to be a very useful technique for non-invasive identification, periodic assessment of the

Fig 2. Coronal (A and B) and transverse (C and D) magnetic resonance images. Dilated right coronary artery arising from the aorta (A, C and D, white arrows) and well-developed vessels in the interventricular septum (D, white arrows) are shown. Anomalous origin of the left coronary artery from the pulmonary artery is also revealed (B, white arrow).

efficacy of therapy, and the follow-up of some coronary artery diseases such as anomalously originating coronary arteries, coronary artery aneurysm in Kawasaki disease, coronary fistulas, and ischemic coronary artery disease.

Acknowledgement

We are grateful to Dr Sachiko Kakitsubata for her suggestions regarding radiology.

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