CASE REPORT

Assessment of Nonviable Myocardium Due to Bland-White-Garland Syndrome Using Contrast-Enhanced MRI

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INTRODUCTION

The left coronary artery originating from the pulmonary trunk is a rare congenital anomaly, known as Bland-White-Garland syndrome (BWGS). Most patients with this anomaly of the coronary artery system die during early childhood (Wesslhoeft et al., 1968). In this case report, we describe an adult patient with nonviable myocardium most likely due to chronic ischemia. To the best of our knowledge, this is the first case report of late myocardial enhancement caused by BWGS detected by cardiovascular magnetic resonance imaging (CMR).

CASE REPORT

An asymptomatic 25-year-old male was referred to our clinic after a systolic murmur was noted during a routine clinical examination. Echocardiography showed a mild mitral regurgitation. The patient developed ST-depression during exercise testing. Exercise myocardial scintigraphy showed hypoperfusion in the anterior wall. Subsequently, a coronary angiography was performed and revealed an enlarged right coronary artery (RCA) and collateral vessels to the left coronary artery (LCA). Selective angiography of the LCA was not possible from the aortic root and even nonselective injection of a contrast agent did not enhance the LCA. Angiography of the pulmonary arteries demonstrated the origin of the LCA from the pulmonary trunk (PT). This anomalous origin of the LCA is known as Bland-White-Garland syndrome.

Contrast-enhanced CMR was performed to analyze the myocardial viability affected by chronic ischemia. Furthermore, MRI was used to demonstrate the anomalous origin of the left coronary artery in a three-dimensional (3D) data set.

Mild hypokinesia of the anterolateral wall and an enlarged left ventricle (EDVI 111 ml/m², ESVI 48 ml/m²) was seen in cine studies [trueFISP, electrocardiogram (ECG)-gated, TR=3.4 ms, TE=1.7 ms, voxel-size 1.4 × 1.1 × 6.0 mm³]. T1-weighted images showed nontransmural late enhancement of the anterior
wall (segmented IR-TurboFLASH-sequence, ECG-gated, TI=220 ms, TE=4.4, voxel-size 1.7 × 1.3 × 6.0 mm³). The enhancement was limited to the endocardial layers (Fig. 1, left panel, arrows). The follow-up MRI examination eight months after cardiac surgery showed the same extent of nonviable myocardium (Fig. 1, right panel).

A short branch of the LCA was depicted using ECG-gated, contrast-enhanced MR-angiography (FLASH-3D, TR=3.1 ms, TE=1.2 ms, voxel-size 1.2 × 0.9 × 2.0 mm³). Figure 2 (multiplanar reconstruction image) shows the LCA originating from the PT. The patient underwent cardiac surgery with proximal ligation of the anomalous LCA at PT. The left internal mammarian artery was used as bypass graft to the LCA.

DISCUSSION

This case report describes for the first time an adult patient with BWGS and documented nonviable myocardium. Because most patients die in infancy, this patient is a rare example of chronic ischemia in a human being. The late myocardial enhancement reflects nonviable myocardium most likely due to chronic ischemia caused by left-to-right shunting and inadequate collateral flow from the RCA in patients with BWGS. The presence of nonviable myocardium could be a risk factor of sudden death. Due to the low incidence of BWGS, this remains hypothetical and is difficult to study systematically. However, the late enhancement technique can identify even small areas of nonviable myocardium (Wagner et al., 2003).

The latest development in scanner systems and sequence techniques could allow CMR to become the only necessary investigation in patients with, or suspected of having, BWGS. Our case report represents a typical clinical cascade of examinations including myocardial scintigraphy and coronary angiography. With the prior knowledge of these examination results, we reduced CMR examination time using a contrast-enhanced MR-angiography and renouncing of a myocardial perfusion study.
However, for a complete examination of BWGS without the typical cascade, we would recommend changing the examination protocol using the following CMR techniques:

1. Typical ECG-gated localizers are used to plan cine long-axis view, cine four-chamber view and cine view of the left ventricular outflow tract.
2. A myocardial perfusion study at rest should be performed based on the localizers in different planes. So far, there exist no guidelines regarding sequence type, or type, dosage, and flow rate of contrast agent. We used a multislice, ECG-gated, saturation recovery, steady-state free precession (SSFP) sequence and 0.05 mmol/kg body weight Gd-DTPA followed by a saline flush, both with an infusion rate of 4 ml/s.
3. We recommend performing a cine short-axis stack encompassing the entire ventricles between the perfusion at rest and under vasodilation (step 5), to allow a minimum of 10 min for the majority of the contrast agent to be washed out. Therefore, ECG-gated cine SSFP sequences are used for analysis of ventricular volumes, myocardial mass, and global and regional function.
4. Noncontrast-enhanced magnetic resonance angiography (MRA) serves to detect coronary artery anomalies. Again, there exist different sequence types. In most cases, the navigator technique with ECG gating is used to prevent respiratory motion artifacts.
5. The myocardial perfusion study as described in step 2 is repeated under adenosine-induced vasodilation (140 mcg/kg/min, started 2–4 min prior to contrast agent bolus). At our institution, we performed two myocardial perfusion studies during vasodilation with a low (0.05 mmol/kg body weight Gd-DTPA) and a high contrast-agent dose (0.15 mmol/kg body weight Gd-DTPA). The second, higher dosage allows for better visual detection of perfusion defects, whereas the smaller dose allows for (semi-)quantitative analysis. Both injections during step 4 result in a typical dosage of 0.2 mmol/kg body weight Gd-DTPA for late enhancement imaging.
6. Late enhancement imaging is started 10–15 min after the contrast agent is administered. The optimal inversion time to null the signal of viable myocardium is determined and ECG-gated, inversion recovery TurboFLASH images are acquired in at least two different imaging planes.

All of the above steps to evaluate patients with or with suspected Bland-White-Garland syndrome can be performed in 40–60 min, providing information on myocardial function, anatomy, perfusion, and viability, as well as an assessment of anomalous coronaries, in one single examination. Cine techniques for evaluation of ventricular motion, function, and volumes and the late enhancement technique for evaluation of nonviable myocardium are considered to be the gold standard (Pohost et al., 2003). Additionally, MRA of the coronary arteries for detection of coronary anomalies is a routinely used clinical approach (Bunce et al., 2003). The detection of ischemia using CMR myocardial perfusion imaging techniques has been highlighted in several studies. Quantitative assessment appears superior as compared to visual analysis (Nagel et al., 2003). However, there is still a lack of multicenter studies and standardized examination protocols.

In conclusion, CMR may become the gold standard to diagnose small areas of nonviable myocardium in patients with Bland-White-Garland syndrome. We introduce a detailed CMR protocol to assess anatomy of coronary anomalies, global and regional cardiac function, volumes, myocardial perfusion, and viability. It is conceivable that young patients with Bland-White-Garland syndrome will benefit particularly from this noninvasive, multimodality CMR approach without radiation.

REFERENCES


