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LETTERS TO THE EDITOR

CMR Evaluation of Cardiac Involvement During the Convalescence of Kawasaki Disease

Kawasaki disease (KD) is a form of vasculitis affecting both the myocardium and coronary arteries. Coronary aneurysms develop in 15% to 25% of untreated patients (1,2). Myocardial inflammation is nearly universal during the acute phase (100% of cases in both post-mortem analysis and myocardial biopsy). However, there are no data about cardiac involvement during the convalescence of KD (1,2). Our aim was to evaluate the heart during the convalescence of KD by cardiac magnetic resonance (CMR). Thirteen KD patients, 6 to 8 years of age who fulfilled the currently used criteria for KD diagnosis, were evaluated by CMR 20 to 40 days after the onset of the disease. Patients were treated with high-dose immunoglobulin within the first 10 days of fever and responded well. Troponin I levels were normal in all except 1 patient, who had ST-segment elevation in II, III, and aVF. Patients were re-evaluated by CMR 3 months later.

Coronary arteries were examined using a 3-dimensional segmented k-space gradient-echo sequence (echo time: 2.1 ms, repetition time: 7.5 ms, flip angle: 30°, slice thickness: 1.5 mm). Inflammation evaluation was performed using short tau inversion recovery T2-weighted analysis, early gadolinium enhancement (EGE), and late gadolinium enhancement (LGE) (contrast-enhanced fast spin echo T1-weighted magnetic resonance, flip angle: 15°, echo time: 1.4 ms, repetition time: 5.5 ms, inversion time: 225 to 275 ms as individually optimized to null myocardial signal). Left ventricular systolic function was evaluated using short-axis steady-state free-precession sequence.

The average length of continuously visualized left main coronary artery with left anterior descending coronary artery (LAD), left circumflex coronary artery, and right coronary artery (RCA) by magnetic resonance angiography was 5.2 ± 0.8 cm, 4.2 ± 1.0 cm, and 9.9 ± 1.2 cm, respectively. The left circumflex coronary artery diameter was 2.01 ± 0.17 mm, the LAD was 3.7 ± 0.2 mm, and the RCA was 3.80 ± 0.09 mm. Coronary artery ectasia (defined as diffuse, uniform luminal dilation between 1.50- to 2.0-fold larger than maximal normal diameter of the respective vessel [1]) was documented in all patients in the LAD and RCA, but not in the left circumflex coronary artery. Discrete, coronary aneurysms (defined as segmental dilation between 1.50- to 2.0-fold larger than maximal normal diameter of the respective vessel [1]) were assessed in 2 patients. An RCA aneurysm (5.5 mm in diameter and 9.5 mm in length) in 1 patient and a LAD aneurysm in another patient (7.5 mm in diameter and 10.5 mm in length) also were identified. Significant stenosis was not detected in any of them. Myocardial inflammation was documented in 6 (46%) of 13 patients. Although statistical evaluation could not be performed because of the small number of patients, in short tau inversion recovery T2-weighted analysis and EGE, the signal ratio was extremely high in KD with inflammation compared with age-matched controls (2.94 ± 0.20 vs. 1.38 ± 0.12 and 28.62 ± 2.48 vs. 2.95 ± 0.21, respectively). The increase in absolute myocardial enhancement in EGE was 160% to 200%. The high values of T2-weighted analysis and EGE in these patients probably were the result of the active inflammatory process early during the course of the disease (examination time: 20 to 25 days after disease onset). The LGE areas were identified in only 3 of 6 patients with myocardial inflammation. A transmural, inferior myocardial infarction in the territory of an RCA aneurysm was assessed in 1 patient (Figs. 1 and 2). Left ventricular ejection fraction was lower in patients with inflammation compared with those without inflammation (left ventricular ejection fraction 49.83 ± 1.16% vs. 62.28 ± 1.70%, $p < 0.001$). No correlation between the presence of coronary aneurysms and myocardial inflammation was documented. CMR re-evaluation of patients 3 months later revealed regression of inflammation in all, whereas coronary ectasia remained unchanged in 5 of 13 patients.

Scarce data exist that are referred to cases with severe left ventricular dysfunction (3–5). Furthermore, there are not

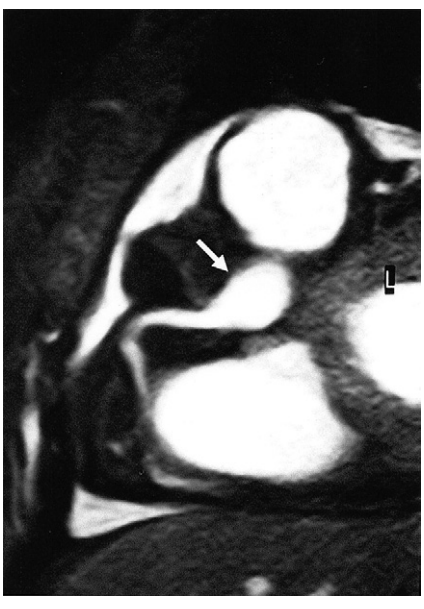

Figure 1. Coronary Magnetic Resonance Angiography Image

Coronary magnetic resonance angiography image showing a right coronary artery aneurysm **(arrow)**.

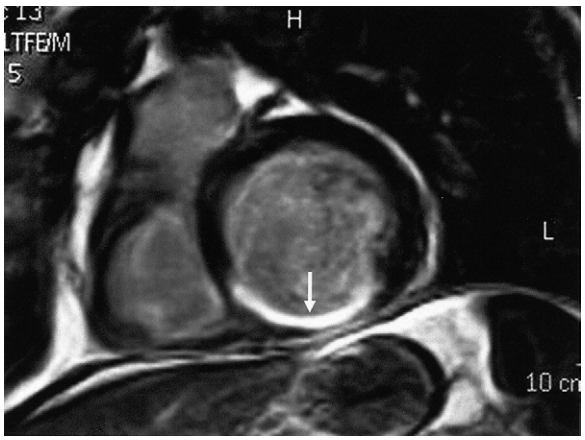

Figure 2. Viability Image

Viability image showing a transmural, inferior myocardial infarction **(arrow)** in the same patient shown in Figure 1.

enough data about CMR for inflammation evaluation in KD. Our findings were in agreement with previous clinical studies supporting the high incidence of myocarditis during stages II and III (days 12 to 25 and days 25 to 40, respectively) and in older patients. In our patients with myocardial inflammation, both T2-weighted analysis and EGE were highly suggestive of inflammatory process. The lack of LGE in one-half of the patients was not a surprising finding. In contrast to viral myocarditis, which is considered to be the result of injury by virus infiltration and the host's immune response, the myocarditis in KD is characterized by inflammatory infiltration from the coronary arteries to the interstitial myocardium, and therefore the myocardial necrosis is not observed frequently, which may explain the lack of LGE and the rapid clinical improvement of our cases (5).

Another important finding was the presence of ectasia, but not aneurysm, in all KD patients. This is probably because the early administration of immunoglobulin decreased the incidence of coronary aneurysms (1). However, myocardial inflammation was a common finding, with rapid recovery in the subsequent 3 months.

The current study has the following limitations: 1) the small number of patients, older than the usual age for KD; 2) infants with severe myocardial disease were not included; 3) long-term CMR follow-up was not available; and 4) myocardial biopsy was not available because of the benign presentation of the disease, which did not allow an invasive procedure.

In conclusion, coronary ectasia and myocardial inflammation are common findings during the convalescence of KD, but usually have a benign prognosis. Myocardial infarction and coronary aneurysms occasionally can be found, although immunoglobulin was administered during the early phase of the disease. A single comprehensive CMR examination applied in KD during convalescence may provide important diagnostic information on myocardial inflammation and coronary anatomy, and thus may be helpful for therapeutic decision making. However, further studies are needed to confirm these findings in larger samples.

Sophie Mavrogeni, MD,* Konstantinos Bratis, MD, Evangelos Karanasios, MD, Dimitris Georgakopoulos, MD, Savas Kaklis, MD, George Varlamis, MD, Genovefa Kolovou, MD, Marouso Douskou, MD, George Papadopoulos, MD

*Onassis Cardiac Surgery Center, 50 Esperou Street, P. Faliro, Athens, 175-61 Greece. *E-mail: soma13@otenet.gr*

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REFERENCES

1. Newburger JW, Takahashi M, Gerber MA, et al. Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. Diagnosis, treatment, and long-term management of Kawasaki disease: a statement for health professionals from the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. Pediatrics 2004;114:1708–33.
2. Mavrogeni S, Papadopoulos G, Douskou M, et al. Magnetic resonance angiography is equivalent to x-ray coronary angiography for the evaluation of the coronary arteries in Kawasaki disease. J Am Coll Cardiol 2004;43:649–52.
3. Mavrogeni SI, Manginas A, Papadakis E, et al. Magnetic resonance angiography, function and viability evaluation in patients with Kawasaki disease. J Cardiovasc Magn Reson 2006;8:493–8.
4. Yoshikawa H, Nomura Y, Masuda K, et al. Four cases of Kawasaki syndrome complicated with myocarditis. Circ J 2006;70:202–5.
5. Shinohara T, Yokoyama T. Clinical study of cardiac pump failure in acute phase of Kawasaki disease. J Pediatr Pract 1999;62:2101–4.

Prosthetic Heart Valve Ring Thrombosis

Despite being mechanical and prosthetic, heart valve rings (HVR) are known to have a lower risk of thrombosis compared with prosthetic mechanical heart valves, thus anticoagulation therapy is only recommended within 3 months after the surgery according to the current guidelines (1). We present 4 cases in which HVR thrombi were obtained by transesophageal echocardiography (TEE) as a novel finding with HVR.

The first case was a 41-year-old woman who had a mitral HVR operation 3 months before her admission with acute atrial fibrillation and who was a candidate for electrical cardioversion. She was referred for TEE to exclude the presence of left atrial (LA) thrombus when we incidentally observed multiple hyperechogenic, mobile thrombus images on the prosthetic mitral ring (Sorin ring, number: 34, Sorin Group, Milan, Italy) (Fig. 1A, Online Video 1A). Her international normalized ratio (INR) was effective (2.6) on admission. As a result of having a history of embolic stroke 1 year previously, the patient was given a low-dose (25 mg), slow-infusion (6 h) tissue-type plasminogen activator (TPA) without bolus administration 2 times (for a total of 50 mg), and as a result, TEE showed an unsatisfactory regression in the thrombi (Fig. 1B, Online Video 1B). We decided to follow up the patient under effective anticoagulation because she refused both the repeat dose of TPA and choice of surgery.

The second case was a 61-year-old woman who had mitral valve replacement (ATS, number: 27, ATS Medical, Inc., Minneapolis, Min-