A 68-year-old white man presented for work-up of recurring atypical resting chest pain in the setting of known coronary artery disease. ECG and chest x-ray on admission can be viewed in Figures 1 and 2. Two years previously, invasive angiography was performed because of unstable angina revealing dilative coronary sclerosis of all coronaries with a proximal right coronary artery stenosis, which was treated with percutaneous intervention by another cardiologist at that time (Figure 3 and online-only Data Supplement Movies I–III).

To evaluate possible progress of coronary artery disease, adenosine stress first-pass perfusion cardiovascular magnetic resonance (CMR) using a 1.5T Magnetom Aera (Siemens Healthcare, Erlangen, Germany) was performed. However, no myocardial ischemia could be detected by CMR in this patient (Figure 4 and online-only Data Supplement Movies IV and V). Late gadolinium enhancement revealed subendocardial infarcts in the inferior and lateral walls (Figure 4).

In the presence of atypical chest pain and 2 subendocardial infarct scars, we performed additional invasive angiography to rule out a false-negative first-pass perfusion CMR study failing to detect flow inhomogeneity as a result of very diffuse coronary disease. We also performed acetylcholine testing for work-up of possible functional coronary disease. Angiography did not reveal any relevant coronary stenosis but confirmed severe dilative coronary sclerosis (Figure 5), which had also been present and reported 2 years before. In addition, an unclear calcified structure (20×17 mm) in proximity to the right coronary artery was detected for the first time (Figure 5). Acetylcholine testing demonstrated epicardial coronary artery spasm with >75% epicardial narrowing in the left as well as in the right coronary artery.
During spasms, the patient reported the same chest pain leading to the actual hospital admission. Chest pain as well as spasms resolved after nitroglycerine, indicating that epicardial coronary spasm was involved in the patient’s current complaints.

Cardiac computed tomography using a 128-slice dual-source Somatom Definition Flash (Siemens Healthcare, Erlangen, Germany) was also performed for evaluation of the unclear calcified cardiac mass seen during invasive angiography. Computed tomography clearly identified the mass as a giant calcified aneurysm of the proximal right coronary artery (31×19×15 mm) with significant intra-aneurysmal thrombus formation (Figure 7). Additional aneurysms could be identified in the distal right coronary and the circumflex artery, indicating that this patient may have suffered from undetected and untreated Kawasaki syndrome during childhood.

Combining all available evidence in this case, one may conclude that this patient suffered Kawasaki syndrome causing multiple calcified (giant) coronary aneurysms and dilative coronary sclerosis. Intra-aneurysmal thrombus formation and subsequent coronary embolism are the most likely mechanism for the 2 subendocardial infarcts in the circumflex and right coronary artery territory, whereas the actual episodes of atypical resting chest pain leading to the current hospital admission are most likely the result of recurring epicardial coronary vasospasm. With this in mind, it remains unclear why Kawasaki syndrome was not already suspected on the basis of the coronary morphology demonstrated by the first invasive angiography performed 2 years previously. However, this diagnosis was not made at that time.

This case is unique in that we do not only visualize the long-term impact of undetected and untreated Kawasaki syndrome on coronary morphology and physiology using a multimodality approach, but also describe coronary vasospasm as a clinically relevant feature in addition to coronary aneurysm formation in this setting for the first time.

Disclosures

None.

Figure 3. First invasive coronary angiography of the left (LCA) (A) and the right (RCA) (B) coronary arteries performed by another cardiologist 2 years previously. Note the proximal RCA stenosis (white arrows) in the setting of diffuse dilative coronary sclerosis (C), which was treated by percutaneous intervention.
Figure 4. Perfusion CMR results can be viewed in the upper panel. (A, B) Note that no perfusion defect is detectable by visual analysis in this patient. Additional post processing to maximize the display of contrast or quantitative perfusion analysis may be helpful in this setting; however, these techniques are not yet widely available to clinicians. The bottom panel (C, D) shows the results of late gadolinium enhancement in the 4-chamber view (4 CH) and the short axis (SAX). White arrows indicate subendocardial infarct scars in the lateral and inferior walls.

Figure 5. Second invasive coronary angiography of the left (LCA) (A) and the right (RCA) (B) coronary arteries. Note severe dilative coronary sclerosis (white arrows) in the entire system. The right image demonstrates an unclear calcified structure (20×17 mm) in proximity to the right coronary artery (white box) (C).
Figure 6. Results of intracoronary acetylcholine testing to identify coronary spasm (A–D). The acetylcholine dose is 80 to 100 μg per vessel (> 200 μg unselective in the left main and 80 μg in the right coronary artery). Note epicardial coronary spasm in the right and the left coronary arteries indicated by white arrows (compare upper to lower panel). ACH indicates acetylcholine.
Figure 7. A, Volume-rendering technique reconstruction of the myocardial surface in left-posterior projection with view on the ramus circumflexus (CX). The white arrow marks an aneurysm of the CX. B, Curved reconstruction of the CX. C, The primary reconstructed transversal data set (the CX aneurysm is marked with a white square). Most notable (D) is a parietal incomplete thrombosis of the aneurysm. D, Volume-rendering technique reconstructed image in right anterior projection with view of the right coronary artery (RCA). This aneurysm could not be assessed satisfactorily by invasive angiogram because of partial thrombosis within the aneurysm (white arrow). The curved reconstruction of the RCA (E) and the primary reconstructed transversal data set prove that the unclear structure shown in Figure 5c is an almost circularly calcified, partially thrombosed RCA aneurysm.
Long-Term Impact of Undetected Kawasaki Syndrome on Coronary Morphology and Physiology
Stefan Grün, Maik Backes, Julia Schumm, Simon Greulich, Peter Ong, Angela Geissler, Udo Sechtem and Heiko Mahrholdt

Circulation. 2012;125:e640-e644
doi: 10.1161/CIRCULATIONAHA.111.076612

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2012 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/125/18/e640

Data Supplement (unedited) at:
http://circ.ahajournals.org/content/suppl/2015/03/02/125.18.e640.DC1

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org/subscriptions/