Biochemical risk assessment and invasive strategies for acute coronary syndromes without ST-segment elevation
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Chapter 11

Ramus Circumflexissimus - a coronary anomaly detected by using computed tomography angiography

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ABSTRACT

A case is presented of a case of L-1 type solitary (left) coronary artery in a female patient with chest pain. It was detected using coronary computed tomography angiography and then confirmed by an invasive coronary angiography. The L-1 subtype was considered to be a benign type. Solitary coronary artery anomalies are rare.
A 52-year-old female was presented to the emergency department with chest pain provoked by emotion. Her ECG and troponin levels were normal. As the echocardiogram was normal, it was decided to perform an exercise test. The test however proved inconclusive. As the likelihood for an NSTE-ACS was estimated to be low, coronary computed tomography angiography (CCTA) was performed, revealing a solitary coronary artery originating from the left sinus of Valsalva. This single coronary artery had a normal take-off and proximal course, dividing into an anterior descending branch and a circumflex branch. After providing blood flow to the posterior descending artery, the superdominant left coronary artery continued into the right posterior atrioventricular groove, supplying marginal branches to the right ventricle and terminating near the right sinus of Valsalva. The right coronary ostium was absent (figure 1). Invasive coronary angiography confirmed these findings (figures 2A and 2B).

Coronary artery anomalies are encountered in 0.6-1.3% of those patients referred for invasive coronary angiography. Contrast-enhanced computed tomography offers additional possibilities to detect coronary anomalies, with the advantage of being non-invasive and providing three-dimensional imaging of the coronary arteries in relation to their surroundings. A rare subset of coronary artery anomalies are the single coronary artery (SCA) anomaly. Isolated SCA anomalies (in the absence of associated cardiovascular anomalies) have been reported with an incidence of 0.04-0.06%. In 1979, A useful classification of SCA anomalies was proposed by Lipton et al, later to be modified by Yamanaka and Hobbs. Our case represents a L-1 type anomaly, originating from the left sinus of Valsalva and following the anatomical course of a normal left coronary artery. The majority of cases of L-1 type SCA described in the literature were detected as a coincidental finding on autopsy or coronary angiography. Typically, its take-off and proximal course do not differ from those of a normal left coronary artery. Kinking or other abnormalities at its origin have not been described. In some cases found during autopsy, a ‘dimple’ was found in the right sinus of Valsalva and in one case, a fibrous chord connected this dimple with the most distal part of the SCA. It is considered that this dimple represents the embryonic bud of the (absent) right coronary artery, which either failed to develop or canalize, or became atretic during an early phase of coronary
artery development. The L-1 type SCA is believed to be a benign anomaly. Like our case, most of the patients reported did not develop symptoms or signs of cardiac dysfunction.\textsuperscript{6-15} However, since the entire heart is supplied by a single coronary artery, the occurrence of atherosclerosis or other pathology in the coronary artery carries an increased risk.\textsuperscript{16-19} Even in the absence of coronary atherosclerosis or thrombosis, ventricular tachycardia and sudden death have both been described in infants as well as in adults with L-1 type SCA. The relationship between the coronary anomaly and the clinical picture of these patients remains unclear and is not supported by evidence of ischemia or by infarction in these reports.\textsuperscript{20-22} Given the relatively small number of known cases and the limited information available on long-term survival in these patients, a definitive statement about the prognosis and optimal treatment of the L-1 type SCA cannot be made. However, in our opinion the risk of cardiovascular events in adult patients with L-1 type SCA will mainly depend upon the development of atherosclerosis in the single coronary artery. Strict regulation of risk factors is therefore of paramount importance as these patients rely on just a single coronary artery.
A rare coronary anomaly detected by CTA

FIGURE 1
Three-dimensional, reconstructed CCTA images showing the solitary left coronary artery, its “ramus circumflexissimus” extending to right ventricular territory.

LAD = left anterior descending; PDA = posterior descending artery; RCxCx = ramus circumflexissimus.
FIGURE 2

Solitary left coronary artery on invasive coronary angiography images.

2A: right anterior oblique view.

2B: caudal left anterior oblique view.
REFERENCES


