Left Main Coronary Artery Arising From the Right Sinus of Valsalva

Ahmed El-Sherief, MD, Richard R. Liberthson, MD, Waleed Ahmed, MD, Carlos A. Rojas, MD, Brian B. Ghoshhajra, MD, MBA, Suhny Abbara, MD, Wilfred S. Mamuya, MD, PhD

Clinical History
A 52-year old man with a history of atypical chest-pain syndrome underwent coronary angiography, which revealed that his left main coronary artery (LM) arose from the right coronary sinus. He was subsequently referred to the MGH Adult Congenital Heart Service for further evaluation, and an ECG-gated coronary CTA was performed to further delineate his congenital coronary anomaly, and to help guide management.

Findings
Cardiac CTA confirmed that his LM arose from his right coronary cusp, and had a separate ostium from the right coronary artery (RCA). The LM had a short sub-pulmonic course with an intramuscular course, with subsequent emergence in the proximal anterior interventricular groove to give rise to the left anterior descending artery (LAD) and left circumflex coronary artery.

Discussion
Congenital coronary artery anomalies are uncommon. Ectopic origin of a coronary artery from the contra-lateral sinus of Valsalva may have important clinical manifestations; most notably exertional sudden cardiac death, particularly in young adults. Ectopic origin of a left coronary artery from the right sinus of Valsalva (RSOV) has been particularly noteworthy for its association with sudden cardiac death, yet not all patients with this anatomical pattern die suddenly.

It is not yet established whether the subgroup of patients with ectopic left coronary artery origin form the RSOV in whom the LM courses within septal musculature as opposed to taking an inter-arterial course are at a lower risk of sudden cardiac death. Our 52 year-old patient, who led a prior physically active lifestyle and has an intramuscular LM course well demonstrated by CTA, is an example of a long-term survivor.

Distinguishing between an inter-arterial course and a myocardial course can be difficult on conventional angiography, and coronary CTA is emerging as the gold-standard in elucidating the origin, the size, the course, and the presence or absence of proximal anatomical or functional obstruction of congenital coronary anomalies. Further observations of these sub-groups of patients by CTA will clarify if indeed there is a differential risk of sudden cardiac death, and the knowledge may have important implications for clinical and management decisions.

REFERENCES
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