Ischemic or Genetic?

Exploring the relationship between an LV aneurysm and anomalous coronary artery

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Introduction

Coronary artery anomalies (CAA) are rare variants that can be diagnosed incidentally on coronary CT angiography (CCTA) during an ischemic evaluation. Reported CAA prevalence varies but may be as high as 7.9% among patients undergoing CCTA.[2] Interestingly, additional congenital variants can coexist with CAA, including congenital left ventricular aneurysm (cLVA). The prevalence of CAA in patients with cLVA is as high as 58%.[4] Ventricular tachycardia or fibrillation occur in 18.4% of patients with cLVA.[5]

Case Presentation

HPI:
76 yo F with HTN, CKD and thyroid disease presented via EMS with acute onset lightheadedness and presyncope while grocery shopping. Evaluation revealed wide complex tachycardia. En route, she was given adenosine 6 mg without effect but later spontaneously converted to sinus bradycardia with PVCs and brief runs of NSVT. She was started on amiodarone and admitted to the cardiology service for further evaluation and treatment. Upon hospital admission she was asymptomatic.

PMHx:
- Grave's disease s/p radiation c/b radiation-induced hypothyroidism
- CKD III
- HTN
- Moderate mitral valve regurgitation

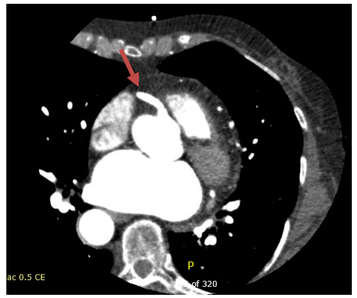
Medications:
- furosemide 40mg daily
- levothyroxine 50mcg daily
- lisinopril 5mg daily
- metoprolol SA 25mg daily
- KCl 20mEq bid

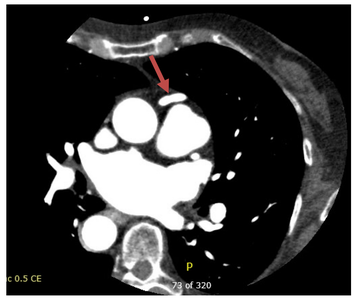
Social Hx:
- Every day smoker – ¼ ppd

Physical Exam:
AF | HR 50 | BP 111/58 | RR 16 | SpO2 100% | BMI 19.7 kg/m²
- General: Healthy-appearing in NAD
- CV: slow rate & regular rhythm, no m/r/g, no carotid bruits, JVP non-elevated, peripheral pulses 2+
- Pulm: CTAB, no crackles
- Abdomen: Soft, ND/NT
- Extremities: No LE edema
- Neuro: A&Ox3, CNII-XII intact
- Psych: Normal mood and affect

Labs:
- Electrolytes wnl
- Cr 1.46
- CBC wnl
- TSH 2.37
- Lipid panel:
  - Total Chol 143
  - TG 103
  - HDL 33
  - LDL 89
- NT-proBNP 622
- hs-Trop trend:
  - 0hr – 58
  - 2hr – 188
  - 3hr – 331
  - 6hr – 388

Imaging





Images 1 (top) **and 2** (bottom)

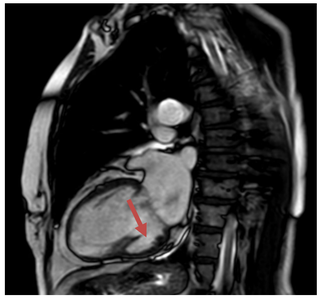
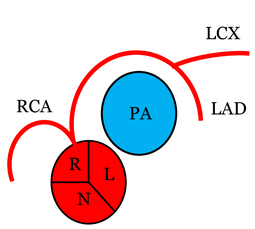


Image 3 (above)



Drawing 1

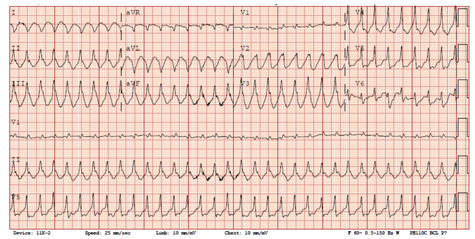


Image 4 (above)

Images 1 and 2 are serial transverse images from a CCTA, demonstrating the single ostium off the right cusp (top) with the left main coursing anterior to the RVOT-PA (lower picture). This course is further illustrated in **Drawing 1**. **Image 3** captures the cLVA at the basal inferolateral wall with late gadolinium enhancement. **Image 4** is the ECG of the patient's ventricular tachycardia.

Hospital Course & Clinical Follow-up

- Patient presented with presyncope and was found to have **wide-complex tachycardia (WCT)** with some characteristics for SVT with aberrancy and some for monomorphic VT.
- Her WCT was resistant to adenosine, and thus a diagnosis of **monomorphic VT** was favored.
- Coronary angiogram demonstrated a **coronary artery anomaly (CAA) with single coronary ostium originating from the right cusp** without obstructive coronary artery disease.
- Cardiac MRI demonstrated a basal inferolateral **left ventricular aneurysm (LVA)** with late gadolinium enhancement (suggestive of myocardial scar).
- Coronary CTA demonstrated the **anomalous coronary artery was coursing anterior to pulmonary artery** (extramural course). Since the LVA was in a non-coronary distribution and non-interarterial CAAs rarely cause ischemia or infarction, the LVA was deemed a **congenital LVA (cLVA)** associated with CAA with VT resulting from its associated myocardial scar.
- An EP study was performed without inducible SVT, evidence of accessory pathway or dual AV node physiology. VT ablation was not performed based on patient's preference.
- Patient underwent placement of a dual-chamber ICD for secondary prevention in the setting of her VT.

Discussion

The patient had a single coronary artery ostium coming off the right cusp of the aorta (Lipton's classification R-IIA), with an anomalous left main coursing anterior to the pulmonary artery before splitting into the LAD and LCX. This course of the CAA portends a lower risk of ischemia and MACE than CAAs that have an interarterial course, which can be compressed between the aorta and pulmonary artery. Given the pre-pulmonic course of her CAA, it was unlikely that the aneurysm of the left ventricle appreciated on cardiac MRI was infarct-related (it was also not in a coronary artery distribution).

Retrospective analyses demonstrate 58% of cLVA patients have associated CAA compared with 6% of a control population ($p<0.001$).[4] Anomalous RCA was the most noted variant among cLVA patients, and our patient's scenario of an anomalous LCA was quite rare.

It was previously estimated that CAA occurred in <1% of the adult population, but increased utilization of invasive and CT coronary angiography has facilitated the discovery of more CAA, leading to an estimated prevalence as high as 7.9%.[2] When considering the risk of ischemia and infarction, one must consider that anomalous LCAs carry higher risk than RCAs, as do CAAs with interarterial course.

Congenital left ventricular aneurysms initially present with arrhythmias in 13% of patients, with 90% of documented arrhythmias being ventricular in nature.[3] It is important to better understand the prevalence and pathophysiology of CAA and cLVA to allow for risk management and primary prevention of fatal arrhythmias or adverse ischemic events.

Genetic testing may facilitate work-up of congenital anomalies. One case report presented a TGFBR1 variant associated with the coexistence of double chamber left ventricle (a cLVA variant), CAA and inducible VT.[1]

Teaching Points

- Depending on their origin and course around the great vessels, coronary artery anomalies can be associated with increased risk of ischemia.
- CAA can coexist with other congenital anomalies, including cLVA.
- Congenital LVA can present with ventricular arrhythmias.

References

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