

Percutaneous Coronary Intervention for Ventricular Fibrillation in the setting of an Anomalous Right Coronary Artery

Danish Saleh¹, Eric Cantey¹, Emily Marogi², Benjamin Freed¹, Bradley Knight¹, Roger de Freitas¹, Ranya Sweis¹, and James Flaherty¹

¹Northwestern University Feinberg School of Medicine

²University of California San Francisco School of Medicine

July 24, 2023

Abstract

We present a case of a young male who presented in urosepsis and developed ventricular fibrillation with cardiac arrest. Work-up revealed a hemodynamically significant anomalous aortic origin of the right-coronary artery. Patient underwent revascularization with percutaneous coronary intervention. Herein, we introduce the case and decision making in our interventional approach.

Title: Percutaneous Coronary Intervention for Ventricular Fibrillation in the setting of an Anomalous Right Coronary Artery

Danish Saleh MD PhD^a, Eric P Cantey MD MS^a, Emily P Marogi MD^b, Benjamin H Freed MD^a, Bradley P Knight MD^a, Roger A de Freitas MD^a, Ranya N Sweis MD MS^a, and James D Flaherty MD^a

^aDepartment of Medicine, Division of Cardiology, Northwestern University Feinberg School of Medicine, Chicago IL. ^bDepartment of Medicine, University of California San Francisco School of Medicine, San Francisco CA.

Keywords/phrases: Anomalous right-coronary artery (aRCA), anomalous coronary artery, ventricular fibrillation, ischemia, percutaneous coronary intervention (PCI)

Words: 1487 (including references and figure legends, excluding abstract and title page)

References 10; Figures 5.

CONSENT STATEMENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

Corresponding author:

Danish Saleh MD, PhD Department of Medicine, Division of Cardiology Feinberg School of Medicine at Northwestern University 676 North St. Clair Street, Arkes Suite 600, Chicago IL 60611 Office: 312-695-0070 Direct: 832-492-5512 Email: danish.saleh@nm.org

ABSTRACT

We present a case of a young male who presented in urosepsis and developed ventricular fibrillation with cardiac arrest. Work-up revealed a hemodynamically significant anomalous aortic origin of the right-coronary artery. Patient underwent revascularization with percutaneous coronary intervention. Herein, we introduce the case and decision making in our interventional approach.

HISTORY OF PRESENTATION

Our patient is a twenty-nine-year-old bedbound male with C4 quadriplegia and a history of atrial fibrillation, who was found to be critically ill with a urinary tract infection, prompting hospital admission. On the fifth day, the patient developed cardiac arrest with ventricular fibrillation. Return of spontaneous circulation (ROSC) was achieved after defibrillation. Patient was supported with electrolyte repletion, anti-arrhythmic therapy, and transvenous pacemaker placement with rapid-ventricular pacing. Targeted-temperature management was enacted per protocol.

PHYSICAL EXAMINATION

On admission, cardiovascular exam was without tachycardia and/or obvious murmurs though the rhythm appeared irregular. Patient had shallow breaths and poor inspiratory effort. Abdomen was diffusely tender, and lower extremities contracted.

PAST MEDICAL HISTORY

Medical history is notable for C4 quadriplegia secondary to a motor-vehicle accident, autonomic-dysfunction, neurogenic-bladder, decubitus ulcers, and recurrent urinary-tract and bone-infections (L ischial tuberosity osteomyelitis) requiring intravenous antibiotics. Patient had atrial fibrillation which was previously managed with propafenone and metoprolol; patient had discontinued these medications a month prior to admission.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for this patient's ventricular fibrillation included myocardial ischemia, structural heart-disease including myocardial scar and/or severe pump dysfunction, congenital and acquired QT-prolonging conditions, severe electrolyte disturbances especially hypokalemia and/or hypomagnesemia, and hemodynamic or metabolic stress in the setting of acute illness.

PERTINENT DATA

At the time of arrest, labs were notable for potassium of 3.0 mEq/L with hemoglobin at baseline (11.7 g/dl). Tele-strips at the time of arrest demonstrated R-on-T phenomenon with Ventricular Fibrillation (Figure 1). Admission high-sensitivity troponin was 12 and initial EKG showed atrial fibrillation alternating with SR and PVCs with a QTc of 431ms (Figure 2a). On admission, two-dimensional transthoracic echocardiography was notable for mildly decreased left ventricular systolic function, and an ejection fraction of 50% with beat-to-beat variation due to frequent ectopy.

INVESTIGATIONS & MANAGEMENT

Post-arrest EKG was notable for ventricular quadrigeminy and QTc of 554ms for sinus beats (Figure 2b) without clear evidence of ischemia. Echocardiogram revealed a depressed ejection-fraction of 21% in the immediate post-arrest period which improved to 40% within one week. Cardiac MRI demonstrated a moderately reduced ejection-fraction of 40% without evidence of myocardial fibrosis. Coronary CT angiogram revealed an anomalous right-coronary artery (aRCA), emerging from the left-main, with significant narrowing between the aortic and pulmonary trunks (Figure 3a, 3b). Calculation of Fractional Flow Reserve (FFR) of the RCA demonstrated a hemodynamically significant anatomic lesion (FFR = 0.51; Figure 3c). Coronary angiogram confirmed significant narrowing of aRCA emerging from the left-main (Figure 4a). Intravascular ultrasound (IVUS) demonstrated a proximal aRCA with extrinsic compression by aortic and pulmonary trunks and ellipse-like cross-section with minimal luminal area of 3.4 mm². Distally, the aRCA had a luminal area of 8.7 mm² (Figure 4b, 4c). The patient underwent PCI to the ostial and proximal RCA with a XienceTM 3.25 x 28mm drug-eluting stent. Post-intervention, IVUS demonstrated rectification of extrinsic compression and recovery of luminal cross-section area (7mm²) associating with TIMI 3 flow on angiogram (Figure 5a, 5b). Post-intervention, the patient was managed with dual-antiplatelet therapy and anticoagulation (for atrial fibrillation and PICC-associated thrombus). Prior to discharge, the patient was transitioned to oral amiodarone and an implantable cardioverter defibrillator (ICD) was placed for secondary prevention. Critical illness and possible osteomyelitis was managed with continued antibiotics.

DISCUSSION

Anomalous aortic origin of the left-coronary artery (AAOLCA) or right-coronary artery (AAORCA) have independently been linked to sudden cardiac death in young individuals [1, 2, 3]. Myocardial ischemia is thought to occur during periods of stress, such as physical activity or periods of heightened metabolic demand, as increased myocardial demand cannot be met due to anatomic obstruction, including vessel compression by adjacent structures (ie: aorta and pulmonary trunk), acute-angle take-off, and/or intramural coronary penetration [4]. Nevertheless, these lesions are observed infrequently and have only been reported in 0.1-1% of births [5].

The optimal approach to ensure durable myocardial perfusion in the setting of a clinical and/or a hemodynamically significant anomalous coronary artery lesion remains unclear. The 2018 AHA/ACC guidelines for the management of congenital heart disease in adults recommend surgery (Class I, Level B-NR) for any AAOCA with myocardial ischemia or AAOLCA without evidence of ischemia (Class IIa/Level C-LD) [6]. Similarly, The American Association for Thoracic Surgeons (AATS) recommend surgery (Class I/Level B) in patients with asymptomatic or symptomatic AAOLCA or symptomatic AAORCA [7]. Surgical approaches include coronary ‘unroofing,’ coronary re-implantation with and/or neo-orifice generation in the correct coronary sinus, pulmonary artery translocation, and coronary-artery bypass grafting (CABG). The AATS advises consideration of PCI for adults with high surgical risk (Class IIb/Level C) [7].

In patients with AAORCA, PCI has been shown to be an effective and low-risk alternative to surgery in limited long-term studies [8, 9]. Indeed, PCI-associated remodeling increases coronary lumen area and features resolution of ischemia [10]. Historically, surgical approaches have been preferred to percutaneous approaches in pediatric patients because of anticipated cardiac/coronary growth and concern for long-term stent-associated complications and/or durability. Although the safety and durability of PCI in the pediatric population has yet to be examined, emerging data supports consideration of PCI in the adults with AAOCA and high surgical risk [7, 8, 9, 10].

FOLLOW UP

There were no further ventricular arrhythmias and/or ICD discharges.

CONCLUSIONS

We suspect that ventricular arrhythmia was the manifestation of ischemia in the setting of congenital flow-limiting stenosis within the right-coronary artery. The ventricular fibrillation threshold was likely lowered by a hypermetabolic state of sepsis, hypotension, and underlying electrolyte abnormalities (hypokalemia). In this young adult with quadriplegia who was a poor candidate for post-surgical rehabilitation, a minimally invasive approach with PCI offered a solution to maintain coronary patency, mitigate myocardial ischemia, and lower the risk of recurrent cardiac arrest.

Funding: None

Acknowledgements: None

Disclosures/Conflicts of Interest: 1. Dr. Knight has received research grants from and has served as a consultant for Abbott, Biotronik, Boston Scientific, and Medtronic. 2. Dr. Sweis is a part of the Speakers’ Bureau with Edwards Lifesciences.

Ethics Statement: We confirm that this manuscript has not been published elsewhere and is not under consideration by another journal. We have removed all identifiable information from this case study for academic and educational purposes. Accordingly, this body of work does not constitute ‘research’ and Institutional Review Board (IRB) approval was not required or obtained. All authors have approved the manuscript and agree with submission to *Clinical Case Reports*.

Author Contributions: D.S. wrote and revised the manuscript and prepared the figures. E.P.C. aided in

manuscript revision and figure preparation. E.P.M. aided in writing the manuscript. B.H.F, B.P.K, R.A.dF, and R.N.S. revised the manuscript. J.D.F. wrote, reviewed, and revised the manuscript.

Data availability statement: Data sharing is not applicable to this article as no new data were created or analyzed in this study.

REFERENCES

- [1] Angelini, Paul. Coronary Artery Anomalies: An Entity in Search of Identity. *Circulation*. March 2007. PMID: 17353457.
- [2] Cheitlin MD, De Castro M, & McAllister HA. Sudden death as a complication of anomalous left coronary origin from the anterior sinus of Valsalva, A not-so-minor congenital anomaly. *Circulation*. 1974. PMID: 4419670.
- [3] Barth CW 3rd and Roberts WC. Left main coronary artery originating from the right sinus of Valsalva and coursing between aorta and pulmonary trunk. *JACC*. 1986. PMID 3944356.
- [4] Taylor AJ, Rogan KM, & Virmani R. Sudden cardiac death associated with isolated congenital coronary artery anomalies. *JACC*. 1992. PMID: 1512344.
- [5] Brothers JA, Frommelt MA, Jaquiss RDB, Myerburg RJ, Fraser CD Jr., & Tweddell JS. Expert consensus guidelines: Anomalous aortic origin of a coronary artery. *The Journal of Thoracic and Cardiovascular Surgery*. June 2017. PMID: 28274557.
- [6] Stout KK, Daniels CJ, Aboulhosn JA, Bozkurt B, Broberg CS, Colman JM, Crumb SR, Dearani JA, Fuller S, Gurvitz M, Khairy P, Landzberg MJ, Saidi A, Valente AM, & Van Hare GF. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American college of cardiology/American heart association task force on clinical practice guidelines. *J Am Coll Cardiol*. 2019. PMID: 30121240.
- [7] Angelini P, Walmsley RP, Libreros A, & Ott DA. Symptomatic anomalous origination of the left coronary artery from the opposite sinus of valsalva. Clinical presentations, diagnosis, and surgical repair. *Tex Heart Inst J*. 2006. PMID: 16878619.
- [8] Darki A, Motiwala A, Bahos L, Lewis BE, Lopez JJ, Steen LH Jr., Mathew V, & Leya FS. Technical success and long-term outcomes after anomalous right coronary artery stenting with cardiac computed tomography angiography correlation. *Catheter Cardiovasc Interv*. Aug 2020. PMID: 31430026.
- [9] Angelini P, Uribe C, Monge J, Tobis JM, Elayda MA, & Willerson JT. Origin of the right coronary artery from the opposite sinus of valsalva in adults: characterization by intravascular ultrasonography at baseline and after stent angioplasty. *Catheter Cardiovasc Interv*. 2015. PMID: 26178792.
- [10] Aubry P, du Fretay XH, Boudvillain O, Degrell P, & ANOCOR Working Group. Place of Angioplasty for Coronary Artery Anomalies with Interarterial Course. *Front. Cardiovasc. Med*. Jan, 2021. PMID: 33614737.

FIGURES LEGENDS

Figure 1 A) A 6-lead telemetry rhythm strip showing atrial fibrillation with natively conducted QRS complexes, aberrantly conducted QRS complexes, PVCs, and the onset of ventricular fibrillation. B) Telemetry strip demonstrating ventricular fibrillation.

Figure 2 A) Admission EKG, QTc ~ 431. B) Post-arrest EKG, QTc ~ 554.

Figure 3 A) CT-Coronary Angiogram demonstrating emergence of an aRCA from the Left Main. B) Dedicated RCA view. C) 3D graphical reconstruction of coronary anatomy with FFR quantitation of coronary flow across proximal aRCA stenosis. FFR of RCA ~ 0.51.

Figure 4 A) Pre-intervention coronary angiogram demonstrating aRCA emerging from the Left Main and region of narrowing. B) Pre-intervention IVUS of proximal aRCA demonstrating ellipse-like luminal compression with luminal area of 3.4mm^2 . Shadowing reflects anatomy of the aortic and pulmonary trunks. C) Pre-intervention IVUS of the aRCA distal to proximal narrowing with luminal area of 8.7mm^2 .

Figure 5 A) Post-intervention coronary angiogram demonstrating Left Main, Left Coronary Circulation, and aRCA with resolution of proximal narrowing. B) Post-intervention IVUS of proximal aRCA post PCI with rectification of luminal integrity and recovery of luminal area (7mm^2). Shadowing reflects anatomy of the aortic and pulmonary trunks.

Hosted file

Figures_Wiley_FINAL.pptx available at <https://authorea.com/users/642500/articles/656321-percutaneous-coronary-intervention-for-ventricular-fibrillation-in-the-setting-of-an-anomalous-right-coronary-artery>