ENDOCARDITIS WITH CORONARY-CAMERAL FISTULA

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Case Vignettes

Background: Coronary-cameral fistulas (CCF) are rare, anomalous connections between a coronary artery and cardiac chamber. It is often asymptomatic in adults and discovered incidentally on cardiac imaging. While acquired cases are reported, most CCFs are congenital anomalies. CCF may predispose patients to the development of infectious endocarditis due to turbulent blood flow.

Case: A 64-year-old male with medical history of hypertension, hypothyroidism, and a recent presumptive diagnosis of vasculitis being treated with oral corticosteroids presented to our institution with three months of progressive fatigue, weight loss, and hematochezia. He was found to have acute on chronic normocytic anemia and admitted for further evaluation. A purpuric rash on his lower extremities prompted evaluation for infective endocarditis with blood cultures ultimately growing *Streptococcus mutans*. Transthoracic echocardiography (TTE) showed a left ventricular ejection fraction of 45 to 50% with a 3 cm aortic valve vegetation and severe aortic insufficiency. Given the size of the vegetation coronary CT angiogram (CCTA) was elected over pre-operative cardiac catheterization. CCTA revealed non-obstructive coronary disease and a coronary-cameral fistula between the left circumflex coronary artery and the junction of the right atrium and coronary sinus with significant dilatation of the left circumflex coronary artery. The patient subsequently underwent successful aortic valve replacement with surgical ligation of the fistula.

Conclusion: Coronary-cameral fistulae present in less than one percent of the population. Often, they are incidental findings due to increasing use of non-invasive imaging techniques, however, congenital CCF have been linked to a variety of complications including infective endocarditis most commonly involving gram positive species. While over half of CCF arise from the right coronary artery, they have been identified in the left coronary system and most commonly drain into either a right-sided chamber or great vessel. The 2018 AHA/ACC consensus statement note that choice of management strategies are controversial and suggest a multidisciplinary approach to determine the role of medical therapy and/or percutaneous or surgical intervention.

- 1. A Vitarelli A, De Curtis G, Conde Y, Colantonio M, Di Benedetto G, Pecce P, De Nardo L, Squillaci E ssessment of congenital coronary artery fistulas by transesophageal color Doppler echocardiography. Am J Med. 2002;113(2):127.
- 2. Gowda ST, Forbes TJ, Singh H, Kovach JA, Prieto L, Latson LA, Turner D. Remodeling and thrombosis following closure of coronary artery fistula with review of management: large distal coronary artery fistula--to close or not to close. Catheter Cardiovasc Interv. 2013;82(1):132. Epub 2013 Feb 14.
- 3. Liberthson RR, Sagar K, Berkoben JP, Weintraub RM, Levine FH. Congenital coronary arteriovenous fistula. Report of 13 patients, review of the literature and delineation of management. Circulation. 1979;59(5):849
- 4. Natarajan A, Khokhar AA, Kirk P, Patel HH, Turner D. Coronary-pulmonary artery fistula: value of 64-MDCT imaging. QJM. 2013;106(1):91. Epub 2011 Dec 20.
- 5. Padfield GJ. A case of coronary cameral fistula. Eur J Echocardiogr. 2009; 10:718–720. doi: 10.1093/ejechocard/jep049
- Spaedy, TJ, Wilensky, RL .Coronary artery fistulas: clinical implications. ACC Current Journal Review. 1994; 3:24.
- 7. Warnes CA, Williams RG, Bashore TM et al., ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (writing committee to develop guidelines on the management of adults with congenital heart disease). Circulation. 2008;118(23):e714.
- 8. Wilde and I. Watt, "Congenital coronary artery fistulae: six new cases with a collective review," Clinical Radiology, vol. 31, no. 3, pp. 301–311, 1980.
- 9. Zhou K, Kong L, Wang Y, Li S, Song L, Wang Z, Wu W, Chen J, Wang Y, Jin Z. Coronary artery fistula in adults: evaluation with dual-source CT coronary angiography. Br J Radiol. 2015 May;88(1049):20140754.