Title: Diffuse Coronary Cameral Fistulas Presenting as a Rare Cause of Coronary Steal Phenomenon

By: Kristen N. Brown, MD

Introduction

Coronary cameral fistulas (CCF) are rare abnormal congenital communications between the coronary arteries (CA) and cardiac chambers. Most commonly CCFs branch from the right CA, and in 90% of cases, CCFs drain into the right cardiac chamber. Most patients with CCFs are asymptomatic. Therefore, CCFs are usually only diagnosed incidentally on coronary angiogram. Classically, the coronary angiogram is the gold standard diagnostic test for diagnosis of CCFs. Management recommendations are very unclear and no treatment guidelines currently exist. Although several reports in the literature have shown success with various types of management approaches including surgical repair, catheter closure, and/or medical management.

Clinical Case

A 65yo woman with a history of COPD, tobacco smoking, hypertension, and reported CAD of LAD on prior heart catheterization who was referred for pre-operative risk stratification with a dobutamine stress echocardiogram. Prior to initiation of the stress test, the patient was noted to angina-free. The patient achieved 92% of maximum Predicted HR. At peak stress, the patient suddenly began to experience severe 9 out of 10 angina that radiated to the jaw. Echocardiographic images demonstrated significant hypokinesis of mid-anterior and mid-anteroseptal wall consistent with left anterior descending (LAD) artery territory ischemia. ECG at peak stress revealed ST elevation in V2 and AVL. 0.4 mg of sublingual nitroglycerin and 5 mg of IV metoprolol were administered without symptom relief. During the recovery period, diffuse hypokinesis was observed except for the basal anterolateral and basal inferolateral segments. Subsequently, an urgent left heart catheterization with coronary angiography was performed. Cardiac catheterization revealed angiographically moderate mid-LAD disease, not flow-limiting with IFR 0.91, nonobstructive left circumflex and right coronary arteries, and diffuse coronary cameral fistulas leading to coronary "steal phenomenon" resulting in myocardial ischemia in the presence of tachycardia explaining the stress echocardiogram findings. Subsequently, perioperative beta blockade and avoidance of intraoperative tachycardia were recommended.

Discussion

This case illustrates a rare yet dangerous and potentially life-threatening presentation of CCF. Typically, clinicians assess for flow-limiting coronary disease as well as significant valvular disease as predictors for worse cardiac outcomes post-operatively. This case, however, demonstrates that the presence of diffuse coronary cameral fistulas can convey the same perioperative cardiac risk as other widely accepted conditions.





Images:

