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When more isn't better: A quadricuspid aortic valve mimicking acute coronary syndrome

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Presentation: A middle-aged male with long standing hypertension and diabetes was initially managed as a case of acute coronary syndrome when he was presented with exertional dyspnea and intermittent angina. Cardiovascular examination revealed a blood pressure of 130/70 mmHg, normal cardiac rate and regular rhythm, a grade 3/5 systolic ejection murmur loudest at the Erb's point, a grade 2/3 early diastolic blowing murmur at the left parasternal border, and a grade 2/5 holosystolic murmur at the apex. Bibasal rales were appreciated.

Imaging Findings: A short-axis view of the great vessels on transthoracic echocardiography revealed an aortic valve with four equally-sized cusps. The presence of marginal calcifications and partial fusion resulted in some restriction in opening during systole, which was confirmed by detection of a significant gradient across the valve. Diastolic malcoaptation was also evident, particularly at the central area, leading to substantial regurgitation. The electrocardiogram showed sinus rhythm with left ventricular hypertrophy. Meanwhile, chest radiography revealed pulmonary congestion and cardiomegaly with left ventricular prominence. A coronary angiogram was performed which demonstrated non-significant coronary artery disease.

Role of Imaging in Patient Care: This case highlights the importance of the prompt use and subsequent correlation of echocardiography with the comprehensive clinical history, physical examination and sound physician judgment in the approach to even the most uncommon cardiac diseases. Discovering them incidentally during surgery or autopsy is now a thing of the past.

Summary/Discussion: The quadricuspid aortic valve is a rare congenital anomaly with a reported prevalence range of only about 0.013~0.043% and requires surgical intervention in more than half of the cases in adulthood due to worsening aortic regurgitation. Previously, it could only be discovered during surgery, aortography or autopsy. Needless to say, echocardiography is an accessible imaging tool to promptly recognize a quadricuspid aortic valve and its associated lesions.