

CLINICAL VIGNETTE

Diagnosis and Management of Acute Coronary Syndrome in a Patient With Dextrocardia

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Case Report

A 58-year-old gentleman with known dextrocardia presented to the Emergency department with chest discomfort. His medical history is significant for a previous myocardial infarction in 1994 requiring percutaneous coronary intervention, hypertension, diabetes, and diverticulitis. His chest pain started 10 days prior to presentation and was intermittent. The pain was right-sided and radiated to this right arm. He presented to his primary care physician, where labs were drawn. The troponin level was elevated and he was immediately referred to the Emergency department.

Vital signs upon arrival were 160/90 mmHg, pulse of 85, and O₂ saturation of 97% on room air. Physical examination revealed mild physical distress and right-sided cardiac auscultation was unremarkable. Electrocardiogram revealed normal sinus rhythm with inverted P-waves in leads I and aVL with reverse R-wave progression in the precordial leads. A right-sided electrocardiogram revealed a previous inferior infarct with non-specific ST and T wave changes.

He was treated with enteric-coated aspirin 325 mg, clopidogrel 600 mg, metoprolol 25 mg, and 2 sublingual nitroglycerin. He was taken urgently to the Cardiac Catheterization Laboratory for coronary angiography.

The coronary arteries were cannulated using the standard diagnostic catheters, however, the radiologic views were altered to

accommodate for the patient's unusual cardiac anatomy. The right coronary artery was noted to have a 90% proximal stenosis with visualized thrombus. The remainder of the coronary vessels had mild to moderate atherosclerotic disease.

Percutaneous coronary intervention was accomplished using a standard right coronary artery guide catheter, a Judkins Right 4 (JR4). Radiologic views were once again modified to accommodate the patient's anomaly, and mostly involved right-anterior oblique views to visualize the proximal portion of the vessel rather than the usual left-anterior oblique views. Intravenous heparin was used for anticoagulation and intravenous eptifibatid was initiated given the thrombus burden. A Pronto thrombectomy extraction catheter (Vascular Solutions Inc., Minneapolis, Minnesota) was used to remove the thrombus and Xience (Abbot Vascular, Santa Clara, California) drug-eluting stent was placed. A left ventriculogram revealed an ejection fraction of 45-50% with inferior wall hypokinesis.

Background

Dextrocardia is a rare congenital cardiac anomaly that affects about 1 in 10,000 births¹. This condition involves an abnormal positioning of the heart in the right hemithorax and can be associated with multiple other cardiac and abdominal abnormalities. Dextrocardia is classified according to the position of the viscera, atria, and great

vessels and includes dextrocardia with situs solitus, dextrocardia with situs inversus, and dextrocardia with situs ambiguus². In situs solitus, the morphological left atrium is to the left of the morphological right atrium, the aorta is on the left, and the abdominal viscera are in its usual location. In contrast, situs inversus has a morphological left atrium that is to the right of the morphological right atrium, a right-sided aorta, a left-sided liver, and a stomach and spleen on the right side. With situs ambiguus, the organization of the atria and visceral organs is variable and inconsistent.

Clinical Aspects of Dextrocardia

The incidence of other congenital cardiac abnormalities in patients with dextrocardia with situs inversus is approximately 3 percent³. The associated cardiac conditions include endocardial cushion defect, single ventricle, transposition of the great vessels, and total anomalous pulmonary venous return. Patients with dextrocardia have a normal life span and low morbidity unless they have an associated rare immotile cilia syndrome known as Kartagener's Syndrome. This is seen in 15% of patients with dextrocardia with situs inversus and presents with repeated chest infections. However, the majority of patients with dextrocardia are usually unaware of their diagnosis until they present with an urgent medical condition. The prevalence of ischemic heart disease is thought to be similar to the general population⁴. Nonetheless, their presentation and diagnosis can present a clinical challenge given the anatomical cardiac anomaly.

Presentation

Although these patients present with chest discomfort, it is right-sided and may radiate to the right arm, shoulder, and jaw. This leads to a complex and often arduous work-up and evaluation in these patients, resulting in a delay in the rapid diagnosis of acute coronary syndrome given the vast differential diagnosis associated with right-sided chest pain.

Diagnosis

The chest X-ray will reveal the diagnosis of dextrocardia, as the heart border will be in the right hemi-thorax. In addition, a 12-lead electrocardiogram (ECG) will reveal inverted P-waves in leads I and aVL. Furthermore, there is progressive loss of the R-wave amplitude in the precordial leads (V1-V6). Patients who present with these ECG characteristics should have a right-sided 12-lead ECG done immediately, especially those in whom acute coronary syndrome is suspected, as the usual 12-lead ECG placement will lead to an erroneous diagnosis. Leads aVL and aVR should be switched to correct the frontal-plane axis and leads V1-V6 should be placed from the left parasternal region to the right mid-axillary line to correct for the anatomic position of the heart.

Management

The management of patients with dextrocardia presenting with acute coronary syndrome is the same as for patients with normal cardiac anatomy and consists of antiplatelet agents such as aspirin and clopidogrel or prasugrel, immediate anticoagulation with heparin, nitroglycerin, and morphine. In those patients referred for coronary catheterization, the cardiac anatomy poses a challenge for the cardiologist. These challenges include catheter selection, coronary engagement, and appropriate radiographic views. Although several different techniques have been described for diagnostic angiography and percutaneous coronary intervention in these patients⁵⁻⁹, it is mostly patient and operator dependent.

Following the immediate management of acute coronary syndrome in dextrocardia patients, aggressive risk factor modification and medical management of coronary artery disease risk factors should be pursued, similar to patients with normal cardiac anatomy. Referral to a congenital heart disease specialist should be considered if other cardiac or visceral abnormalities are suspected.

Prognosis

The prognosis of dextrocardia patients presenting with acute coronary syndromes is similar to normally oriented hearts. Nonetheless, any delay in the diagnosis and treatment of acute coronary syndromes due to different chest-pain presentation and atypical ECG placement, may lead to worsening outcomes. It is imperative that a high index of clinical suspicion leads a clinician to a rapid diagnosis and subsequent appropriate management for this infrequent cardiac anomaly.

Conclusion

Dextrocardia is a rare congenital cardiac anomaly that involves a rightward orientation of the heart. These patients have a similar incidence of coronary artery disease, although their acute coronary syndrome presentation may pose a diagnostic challenge. Both immediate and long-term management of dextrocardia patients presenting with acute coronary syndromes is similar to the general population.

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