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Ebstein's Anomaly Assessed by Cardiac MRI

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Abstract:

Patients with congenital heart disease present a challenge for imagers, since it requires a profound knowledge of the morphologic and functional characteristics of a broad range of congenital heart defects. Ebstein's anomaly is an uncommon congenital abnormality of tricuspid valve characterized by dysplasia and downward displacement of the septal and posterior leaflets. We present a case of a young male who presented for further cardiac work-up to evaluate for suspected Ebstein's anomaly after prior history of radiofrequency catheter ablation for Wolff-Parkinson-White syndrome. The case highlights the diagnostic utility of cardiac magnetic resonance imaging in identifying the anomaly.

Key words: Ebstein Anomaly, Congenital Heart Defects, Wolff-Parkinson-White Syndrome, Magnetic Resonance Imaging.

Introduction

Cardiac magnetic resonance imaging provides a sensitive non-invasive imaging tool to accurately delineate pathophysiologic changes in congenital heart disease for both morphological and functional aspects. It can be reliably used for initial diagnosis as well as for follow-up.

Case Report

A 31 year-old man was referred to our hospital for cardiac work-up of suspected Ebstein's anomaly (EA) from an outside hospital. He was asymptomatic on presentation and had a past history of radiofrequency catheter ablation for Wolff-Parkinson-White syndrome (WPW).

He underwent cardiac magnetic resonance imaging (CMRI) which demonstrated an apically displaced septal leaflet of the tricuspid valve (TV) (23 mm or 11 mm/m^2) along with atrialized portion of the right ventricle (RV) with dilated right atrium [Fig.1]. There was no atrial or ventricular septal defect or evidence of intra-cardiac shunting on phase contrast imaging. The functional RV had normal size and function and there was moderate TV regurgitation. CMRI findings were consistent with EA and thus helped in confirming the diagnosis.

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Discussion

EA is a rare malformation of the TV that occurs in approximately 0.5%-1% of patients with congenital heart disease [1]. It is characterized by varying degrees of dysplasia and downward displacement of the tricuspid valve leaflets into the RV with the anomalous valve often being regurgitant. These anatomic defects divide the RV into two components: a thin walled proximal portion of the RV that becomes atrialized and enlarged; and a more distal trabeculated component constituting the functional right ventricle [1,2]. It is frequently accompanied by other abnormalities such as atrial or ventricular septal defect, patent foramen ovale, pulmonary stenosis or atresia, and myocardial abnormalities.

EA is the most commonly occurring congenital defect associated with WPW syndrome [3]. It has been shown that patients with EA who undergo accessory pathway ablation for WPW syndrome have fewer hospitalizations and atrial fibrillation post-procedure [3].

Compared to other imaging modalities, CMRI is well suited for the evaluation of patients with EA as it has excellent spatial resolution and an ability to clearly distinguish the blood-myocardial border. It enables accurate physiological assessment of ventricular volume and function, extent of tricuspid regurgitation, leaflet size and location, annulus and atrial size as well as other associated cardiac defects [4,5].

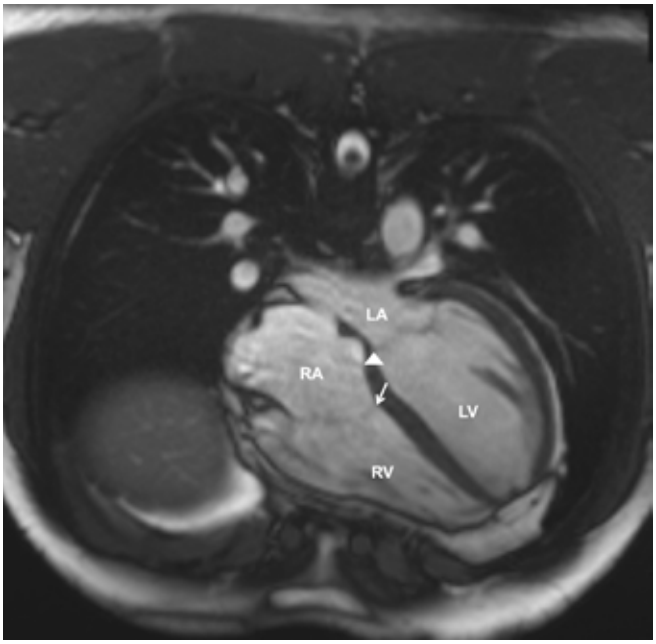

Fig.1: Apically displaced septal leaflet of the tricuspid valve along with atrialized portion of right ventricle (arrow). RA: right atrium; RV: right ventricle; LA: left atrium; LV: left ventricle.

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