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Case Reports

Congenitally unguarded tricuspid valve orifice with right ventricular apical isolation in an adult

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ABSTRACT

Congenitally unguarded tricuspid valve (TV) orifice, a variant of TV dysplasia, is a rare malformation with protean manifestations. This report describes a symptomatic adult male with gross right heart failure and atrial fibrillation, who was found to have an unguarded TV orifice with isolation of the trabecular apical cavity of the right ventricle (RV) and muscular ridges separating outflow tract (forme-fruste of the double-chambered RV). The right ventricular outflow tract remained patent.

1. Introduction

Unguarded tricuspid valve (TV) with well-developed right ventricle (RV) and the patent outflow tract constitute a rare congenital anomaly with a few antemortem case reports.[1–11] It is a variety of TV dysplasia, in which there is partial or complete agenesis of the TV tissue.[12] The leaflets are normally inserted on the ring and there is variable dysplasia of chordae tendinae and papillary muscles. Unguarded TV orifice with patent right ventricular outflow tract and dilated RV in adults is very uncommon and needs to be differentiated from the more common entity – Ebstein anomaly of the TV. The hallmarks of poor prognosis, regardless of the valvar displacement are lung hypoplasia, right atrial dilatation, and the relative hypoplasia of the pulmonary trunk. There are two types of non-Ebstein TV dysplasia: one is a more frequent type having dysplasia of TV with a small annulus, underdeveloped RV with a hypoplastic cavity and a hypertrophic wall; the other type has severe dysplasia of TV and dilatation of RV, right atrium (RA), and right atrioventricular junction with thinning of the RV wall. The latter type is associated with patent outflow tract. RV in TV dysplasia can be underdeveloped, normally developed or dilated and hypokinetic. The last variety is associated with congenitally unguarded TV orifice. The difference between the Ebstein anomaly and the unguarded TV orifice is best demonstrated by examining the mural leaflet of the valve, which is absent or markedly underdeveloped, when the orifice is unguarded but displaced in association with Ebstein's malformation. Ebstein's anomaly is confidently ruled out with comprehensive echocardiography by establishing (1) absence of significant apical displacement of the septal TV leaflet (≥8 mm/m^2) and (2) lack of a redundant, elongated, anterior TV leaflet. The clinical presentation is usually in early childhood with cyanosis and/or congestive heart failure. There are a few patients, who decompensate during adult life with

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right ventricular failure and tricuspid regurgitation.[8–10] The advent of echocardiography has resulted in a significant number of such patients being diagnosed. Some of these patients may present with atrial fibrillation.[8,9] This report describes a symptomatic young man presenting with gross right heart failure and atrial fibrillation who on echocardiography was detected to have unguarded but large TV orifice, hugely dilated RA and the RV with isolation of the apical trabeculated part of the right ventricular cavity with a fibromuscular ridge.

2. Case report

A 32-year-old gentleman presented with exertional dyspnea, fatigue, and pre-syncope of three years' duration. Physical examination revealed a thin-built young man with supine blood pressure 102/82 mmHg, pulse rate 102/min; the rhythm was irregular, and jugular venous pressure was markedly elevated with prominent V waves and neck nodding associated with bilateral jugular vein distension with each heart beat. There was significant non-tender pulsatile hepatomegaly. Precordial examination revealed quiet precordium on palpation, faint heart sounds with wide variably split second sound, and a 2/6 systolic murmur. Hematological parameters and plasma biochemistry were normal. A 12-lead electrocardiogram revealed right axis deviation, atrial fibrillation with an average ventricular rate of 102/min, complete right bundle branch block and non-specific ST-T changes (Fig. 1). A plain chest skiagram showed cardiomegaly (cardiothoracic ratio ~70%) and oligemic lung fields (Fig. 1).

Two-dimensional echocardiography showed small left atrium and the left ventricle, markedly dilated right heart chambers, paradoxical ventricular septal motion, interatrial septum curved to the left, dilated inferior vena cava and hepatic veins, enlarged tricuspid annulus (48 mm), rudimentary septal and anterior tricuspid leaflet with a nodular mass attached to the vestigial anterior leaflet (Fig. 2). Septal leaflet of the TV was rudimentary but normally inserted. Posterior tricuspid leaflet and the papillary muscles were not visible. The right ventricular outflow tract was dilated with normal pulmonary arteries. Doppler interrogation of the right ventricular inflow and outflow showed low-velocity to-and-fro flow with a peak velocity of <1 m/s (Fig. 3).

In the right ventricular cavity, a fibromuscular ridge separated distal apical portion of the cavity from the right ventricular inflow. The apical portion was highly trabeculated and on color flow interrogation showed no communication with the proximal cavity (Fig. 4). Well-defined septal and parietal muscle bundles separated the right ventricular outflow tract. This segment showed dynamic systolic narrowing without any pressure gradients (Fig. 5).

The patient is awaiting total cavo-pulmonary anastomosis.

3. Discussion

Congenitally unguarded tricuspid orifice with patent right ventricular outflow tract is a rare anomaly with <50 cases reported in the literature with antemortem diagnosis.[1–11] Isolated unguarded tricuspid orifice with no other congenital abnormality has been reported in a few patients so far.[1,8–10]

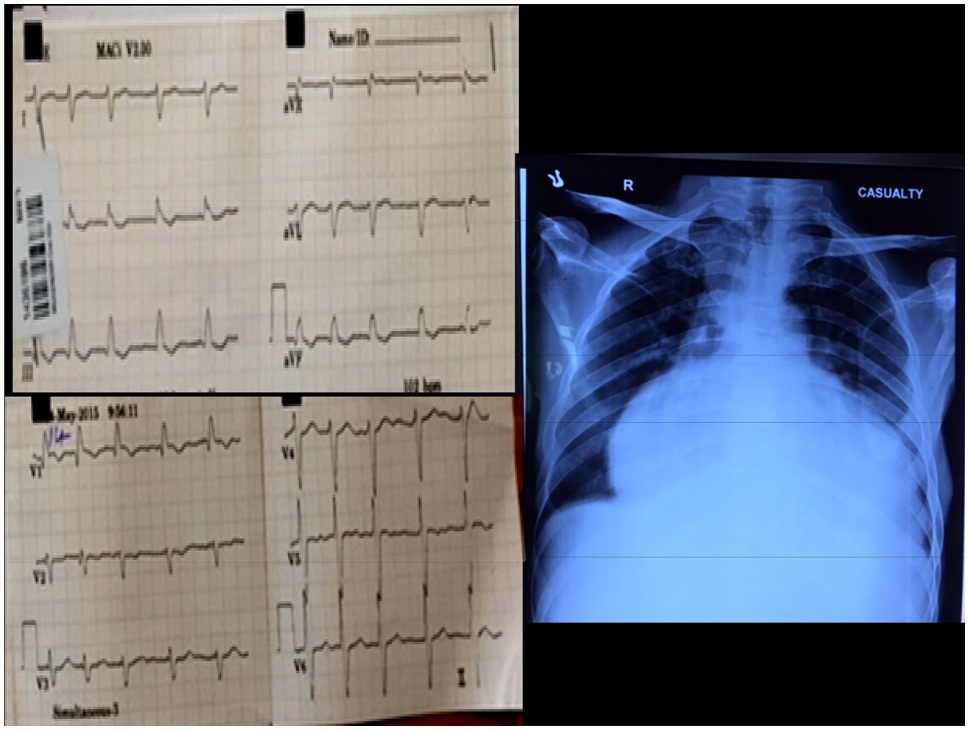

Fig. 1 – The 12-lead ECG in the left panel shows atrial fibrillation, right axis deviation, right bundle branch block and non-specific ST-T changes. Right panel shows chest skiagram in the antero-posterior view. Note globular cardiomegaly with pulmonary oligemia.

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