Journal of Coronary Heart Diseases

Charan et al., J Coron Heart Dis 2019, 3:1

Case Report Open Access

Diagnosis and Management of Isolated Tricuspid Regurgitation: An Enigma

Charan RKV*, Prakash Sanzgiri, Vedanti Shingare and Vidya Suratkal

Department of Clinical and Interventional Cardiology, Lilavati Hospital and Research Centre, Bandra, Mumbai, India

Abstract

Secondary or functional Mitral Regurgitation (MR) is well documented. Long-standing MR, Pulmonary Hypertension (PH) or myocardial disease usually leads to Secondary Tricuspid Regurgitation (STR). The following case of chronic Left Ventricular (LV) ischemia and the resultant myocardial scarring causing isolated Tricuspid Regurgitation (TR) without left heart dysfunction has not yet been reported.

Keywords: Functional tricuspid regurgitation; Left heart dysfunction; Valvular heart disease

Abbreviations: MR: Mitral Regurgitation; TR: Tricuspid Regurgitation; PH: Pulmonary Hypertension; CABS: Coronary Artery Bypass Surgery; LVEF: Left Ventricular Ejection Fraction; RWMA: Regional Wall Motion Abnormality; CMRI: Cardiac Magnetic Resonance Imaging

Introduction

Tricuspid regurgitation (TR) is an important clinical problem because it is frequent and carries a poor prognosis when it is left uncorrected [1,2]. Mild TR is common and usually is benign. However there is still a lack of awareness of tricuspid disease in the medical community. The recent ESC/EACTS guidelines [3] stress the importance of the awareness of tricuspid disease especially in patients with significant mitral valve disease. TR occurs mainly from tricuspid annular dilation, which can result from left-sided heart failure from myocardial or valvular causes, right ventricular volume and pressure overload, or dilation of cardiac chambers [4,5]. The patients with severe TR, especially secondary TR, are often at an advanced stage of the disease and require a comprehensive cardiac and non-cardiac evaluation. If untreated at the time of surgical mitral valve repair, significant residual TR negatively impacts perioperative outcomes, functional class, and survival [6-8].

Tricuspid Valve (TV) and its associated diseases have been ignored for a long time [9]. The incidence of tricuspid insufficiency associated with left valvular disease appears to be increasing and ranges from 8% to 35% of cases [9]. Though it is frequently related to rheumatic process affecting other valves, one should also be aware of the secondary or functional causes affecting the TV apparatus. Moreover, there has been no mention of a case of isolated functional TR purely attributed to an ischaemic cause without concomitant left heart dysfunction. This case describes the temporal association of ischemia and TV disease, the mechanism involved, diagnosis and various treatment strategies.

Description of Case

73-year-old, non-diabetic, non-hypertensive male presented with fatigue and progressive swelling in both feet over the period of the past 6 months. There was no angina, syncope, orthopnea or Paroxysmal Nocturnal Dyspnea (PND). His past history revealed an anterior wall ST-Elevation Myocardial Infarction (STEMI) 14 years back. He was first thrombolysed with Streptokinase (STK) at a peripheral center, and 4 hrs later underwent rescue coronary angioplasty to the culprit's vessel, the Left Anterior Descending (LAD) artery with a Cypher 3 mm × 12 mm Drug-Eluted Stent (DES). The coronary angiogram revealed severe triple vessel disease. 2D-ECHO showed hypokinesia of the anterior wall but preserved wall thickness and LV-Ejection Fraction (EF) of 45%, structurally normal valves with no Mitral Regurgitation (MR) or TR. He was discharged on aspirin (75 mg/day), clopidogrel (75 mg/day) and atorvastatin (40 mg/day).

Subsequently, after six weeks, he was advised to undergo coronary bypass surgery to enable total revascularisation. On-pump Coronary Artery Bypass Surgery (CABS) was successful with a LIMA-RIMA Y-grafting to Left Anterior Descending (LAD), Left circumflex (Lcx) and Right Coronary Arteries (RCA).

The patient was asymptomatic for the next 6 years but presented with pain in the abdomen, icterus, and bilious vomiting. CT abdomen revealed the cause of obstructive jaundice to be a tumor in the peri-ampullary region. Transduodenal ampullectomy was performed. The histology revealed non-malignant villous adenoma with mild atypia. 2D-ECHO showed normal left Ventricular (LV) and Right Ventricular (RV) systolic function with Left Ventricular Ejection Fraction (LVEF) of 55% to 60% and no Regional Wall Motion Abnormality (RWMA). Structurally normal valves and a trivial TR. He was later discharged and was asymptomatic until the present symptoms 6 months back.

On examination, Jugular Venous Pressure (JVP) was raised with a prominent 'V' wave and positive hepato-jugular reflux. On auscultation, a high pitched holosystolic murmur of grade III/VI at the left lower sternal border radiating to the right lower sternal border was audible. The murmur is accentuated on inspiration and by passive leg raising. S3 gallop was present. ECG showed AF with a controlled heart rate (80 bpm) (Figure 1). Additional tests done showed haemoglobin to be 13 gm/dl and NT-proBNP of 3047 pg/mL (normal value is less than 125 pg/ml).

2D-ECHO (Figure 2a and 2b) and later 3D-ECHO (Figure 2c and 2d) was performed to assess valvular morphology, which showed normal LV size and LVEF function (50%) and no RWMA. LA volume was normal, structurally normal MV apparatus and aortic valve. RA was dilated with an area of 28 cm^2 (normal ≤ 18 cm^2) with inadequate coaptation of TV leaflets with a tenting area of 1.9 cm^2 (normally less

***Corresponding author:** Charan RKV, Department of Clinical and Interventional Cardiology, Lilavati Hospital and Research Centre, Bandra, Mumbai, India, E-mail: chrnr@rediffmail.com

Received May 20, 2019; **Accepted** June 03, 2019; **Published** June 06, 2019

Citation: Charan RKV, Sanzgiri P, Shingare V, Suratkal V (2019) Diagnosis and Management of Isolated Tricuspid Regurgitation: An Enigma. J Coron Heart Dis 3: 113

than 1.6 cm^2). TV annular diameter (in diastole) was 4.0 cm (normal ≤ 2.8 cm), RV ejection fraction was 45%. Pulmonary artery pressure was normal. No thrombus in any of the heart chambers. No myocardial scarring or RV endocardial thickening. Doppler study showed moderate TR (Jet area/RA area=25%), vena contact width of 6 mm and mild MR. Inferior vena cava was dilated (2.1 cm) with less than 50% collapse on inspiration.

Urinary 5-Hydroxyindoleacetic Acid (5-HIAA) was also done to rule out carcinoid syndrome, given the history of gastrointestinal tumor in the past. MRCP of the abdomen was done, which did not reveal any new growth in the peri-ampullary region or metastatic deposits elsewhere.

Cardiac MRI with gadolinium contrast (Figure 3) showed a late enhancement, most likely due to a trans-myocardial scar involving basal inferoseptal and mid-infero-septal walls extending into the mid-inferior wall with dyskinetic septal movement. The posterior TV leaflet was tethered to this scar resulting in its restriction with incomplete coaptation, leading to Tricuspid Regurgitation. RV end-diastolic volume was 145 ml/m^2 and RV end-systolic area was 15 cm^2. No endocardial thickening of RV or structural abnormalities of chordae or chordal insertion points was observed. The patient was treated with intravenous Furosemide. He is currently on regular follow-up and is asymptomatic at present.

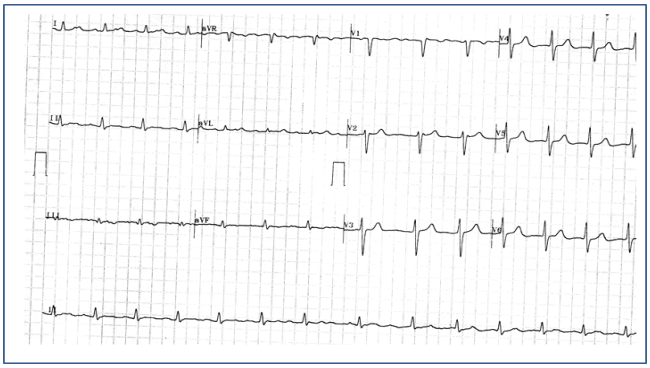

Figure 1: Figure showing atrial fibrillation with controlled heart rate (80 bpm).

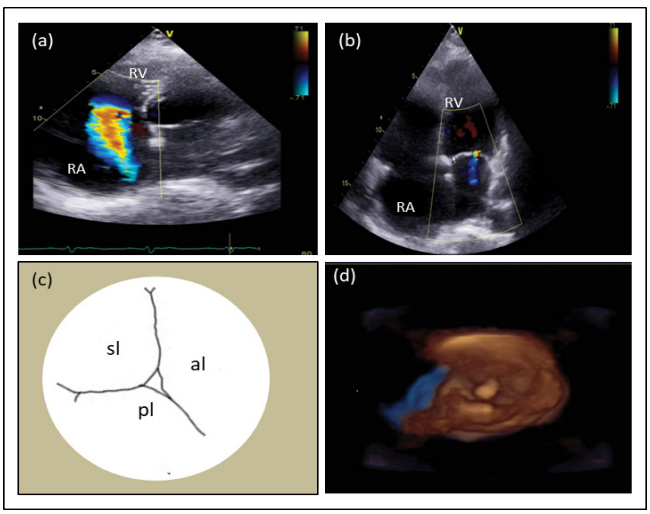

Figure 2: 2D Echocardiographic (colour Doppler) assessment of tricuspid valve (a): showing moderate tricuspid regurgitation; (b): Trivial mitral regurgitation; (c): Normal tricuspid valve (En-face view); and (d): 3D Echocardiographic view shows a tethered posterior tricuspid leaflet (RA: right atrium; RV: right ventricle, pl: posterior leaflet, al: anterior leaflet, sl: septal leaflet).

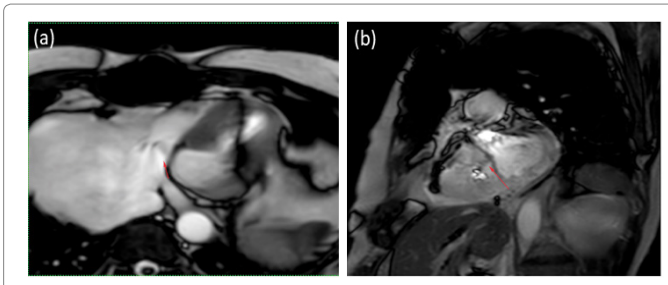

Figure 3: (a): Cardiac MRI showing trans-septal scar (red arrow) (b): leading to isolated tricuspid regurgitation due to tethering of posterior tricuspid leaflet.

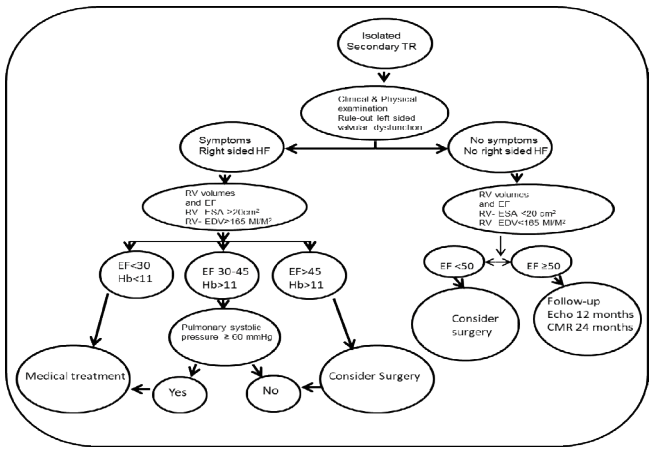

Figure 4: Flow chart showing approach to isolated Tricuspid Regurgitation (TR). (EF: Ejection Fraction; HF: Heart Failure; RV-ESA: Right ventricular End Systolic Area; RV-EDA: Right Ventricular End Diastolic Area; Hb: Hemoglobin).

Discussion

TR was first described by King [10], who showed that distension of the RV with water induced considerable TV regurgitation into the right atrium. Long-standing TR causes progressive dilatation of Right Atrium, inferior vena cava, hepatic veins, and coronary sinus. TR leads to progressive RV volume overload and increased RV diastolic pressure, the interventricular septal shift towards the left ventricle and reduction in cardiac output [11]. This remodeling is also known to cause displacement of papillary muscles and valvular tenting leading to poor TV coaptation. This vicious cycle progressively worsens TR leading to RV dilatation and Heart Failure (HF).

Primary TV disease which accounts for 10% of all cases of TV regurgitation may be seen in congenital conditions such as Ebstein's anomaly and atrioventricular canal defects [12]. The primary TV can also be seen in acquired conditions include endocarditis, rheumatic disease, carcinoid syndrome, Marfan syndrome, radiation exposure or flail leaflet.

Secondary TV disease accounts for 80% of cases result from annular dilation or leaflet tethering leading to poor coaptation of TV leaflets with resultant TR [13]. Fukuda et al. [14] have shown septal leaflet tethering in patients with secondary TR who have normal pulmonary artery pressures. Change in the geometry of the RV, and the consequent papillary muscle displacement is the critical factor in the pathophysiology of secondary TR [15].

Echocardiographic assessment of the TV is often challenging due to the unfavorable retrosternal position of the valve and the inability to simultaneously visualize all three leaflets in standard transthoracic views. However, Cardiac Magnetic Resonance Imaging (CMRI) gives a definitive diagnosis. The presence of thinning with the dyskinetic movement of the inter-ventricular septum and Late Gadolinium Enhancement (LGE) often gives information regarding the etiopathogenesis [16]. Evidence of tethering of posterior leaflet of TV to scarred septal myocardium and mal-apposition leading to progressive TR clinched the diagnosis in our case. Many guidelines still recommend TV surgery or repair for patients with severe TR at the time of left valve surgery (Figure 4), preferentially before the onset of significant RV dysfunction and anemia.

Conclusion

Functional or secondary MR causing restriction of leaflet closure can result in the dilation of the mitral annulus, papillary muscles displacement and reduced closing force of leaflets without primary valve leaflet pathology. However, there has been no mention in medical literature, of Isolated Functional Tricuspid regurgitation purely attributed to an ischaemic scar as a cause, without concomitant left heart dysfunction

This rare case also highlights the benefits of using cardiac MRI in the evaluation of unusual valvular cases which defy logical assessment. CMRI can provide comprehensive morphological and functional information on TV with good spatiotemporal resolution and multiplanar imaging capabilities without the use of ionizing radiation.

Conflict of Interest

The authors declare no conflict of interest.

Acknowledgment

The authors are thankful to the patient for his consenting us to publish this report. We are also thankful to the Cardiology ICU staff of Lilavati Hospital and Research Center for their support during the study.

References

1. Dreyfus GD, Martin RP, Chan KM, Dulguerov F, Alexandrescu C (2015) Functional tricuspid regurgitation: a need to revise our understanding. J Am Coll Cardiol 65: 2331-2336.
2. Vahanian A, Brochet E, Juliard JM (2018) Guidelines recommendations on the treatment of tricuspid regurgitation. Where are we and where do we go with transcatheter valve intervention. Front Cardiovasc Med 5: 37.
3. Baumgartner H, Falk V, Bax JJ, De Bonis M, Hamm C, et al. (2017) ESC/EACTS Guidelines for the management of valvular heart disease. Eur Heart J 38: 2739-2791.
4. Nishimura RA, Otto CM, Bonow RO (2014) AHA/ACC guideline for the management of patients with valvular heart disease: A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol 63: e57-185.
5. Latib A, Mangieri A (2017) Transcatheter tricuspid valve repair: New valve, new opportunities, new challenges. J Am Coll Cardiol 69: 1807-1810.
6. Rogers JH, Bolling SF (2009). The tricuspid valve: Current perspective and evolving management of tricuspid regurgitation. Circulation 119: 2718-2725.
7. Topilsky Y, Nkomo VT, Vatury O (2014) Clinical outcome of isolated tricuspid regurgitation. JACC Cardiovasc Imaging 7: 1185-1194.
8. Arsalan M, Walther T, Smith RL, Grayburn PA (2017) Tricuspid regurgitation diagnosis and treatment. Eur Heart J 38: 634-638.
9. Mas TP, Palomares RJF, Antunes MJ (2015) Secondary tricuspid valve regurgitation: A forgotten entity. Heart 101: 1840-1848.
10. King TW (1837) Guy's Hosp. Rep 2: 104.
11. Fender EA, Zack CJ, Nishimura RA (2018) Isolated tricuspid regurgitation: outcomes and therapeutic interventions. Heart 104: 1-109.
12. Mutlak D, Lessick J, Reisner SA, Aronson D, Dabbah S, et al. (2007) Echocardiography-based spectrum of severe tricuspid regurgitation: the frequency of apparently idiopathic tricuspid regurgitation. J Am Soc Echocardiogr 20: 405-408.
13. Huttina O, Voilliot D, Mandryb D, Vennera C, Juillière Y, et al. (2016) All you need to know about the tricuspid valve: Tricuspid valve imaging and tricuspid regurgitation analysis. Arch Cardiovasc Dis 109: 67-80.
14. Fukuda S, Saracino G, Matsumura Y, Daimon M, Tran H, et al. (2006) Three-dimensional geometry of the tricuspid annulus in healthy subjects and in patients with functional tricuspid regurgitation: A real-time, 3-dimensional echocardiographic study. Circulation 114: 1492-1498.
15. Prihadi EA (2018) Tricuspid valve regurgitation: no longer the "forgotten valve". European Soc Cardiol 16: 1-5.
16. Satoh H, Sano M, Suwa K, Saitoh T, Nobuhara M, et al. (2014) Distribution of late gadolinium enhancement in various types of cardiomyopathies: Significance in the differential diagnosis, clinical features and prognosis. World J Cardiol 6: 585-601.