MED-PHOENIX: JOURNAL OF NATIONAL MEDICAL COLLEGE

CASE REPORT

A RARE CASE REPORT ON RHEUMATIC ORIGIN OF SEVERE TRICUSPID STENOSIS

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Date of Submission : May 10, 2021
Date of Revision : Jun 10, 2021
Date of Acceptance : Jun 30, 2021
Date of Publication : July 19, 2021

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Citation:
Laudari S, Tiwari KK, Adhayaru S, Subedi A. A Rare Case report on Rheumatic Origin of Severe Tricuspid stenosis. Medphoenix. 2021;6(1):56-58

DOI:https://doi.org/10.3126/medphoenix.v6i1.37022

Conflict of interest: None, ***Funding:*** None

Publisher: *National Medical College Pvt. Ltd.*
MedPhoenix - Journal of National Medical College (JNMC); 2021,6(1), available at www.jnmc.com.np

ISSN:2631-1992 (Online); ISSN:2392-425X (Print)

ABSTRACT

Tricuspid stenosis is a very rare valvular disease due to narrowing of the orifice of the tricuspid valve of the heart. It is usually of rheumatic origin which is accompanied by other valvular lesions. Other causes of tricuspid stenosis include carcinoid syndrome, endocarditis, endomyocardial fibrosis, lupus erythematosus, right atrial myxoma, drug induced and congenital tricuspid atresia. Here we report a patient who had undergone percutaneous transluminal mitral commissurotomy (PTMC) followed by mitral restenosis with Severe Tricuspid Stenosis with Severe Tricuspid Regurgitation.

Keywords: *Tricuspid stenosis; Rheumatic; Mitral stenosis*

INTRODUCTION

Tricuspid Stenosis(TS) accounts for about 2.4% of all cases of organic tricuspid valve disease and is mostly seen in young women. [1] The congenital form of the disease has a slightly higher male predominance. [2] Around 15 % patients with rheumatic heart disease at autopsy show evidence of TS. But it is clinically significant in less than 5% of cases. [3]

Tricuspid Stenosis is almost always rheumatic in origin. Most patients with rheumatic tricuspid valve disease present with Tricuspid Regurgitation (TR) or a combination of TS and TR. Isolated rheumatic TS is uncommon, and this lesion generally accompanies mitral valve disease. Other causes of tricuspid stenosis include carcinoid syndrome, endocarditis, endomyocardial fibrosis, lupus erythematosus, congenital tricuspid atresia, extracardiac tumours, pacemaker lead and fusion of implantable cardioverter defibrillator leading to sub-valvular structures damage. [4,5]

With the exceptions of congenital causes or active infective endocarditis, tricuspid stenosis takes years to develop. [2] Rheumatic tricuspid disease is characterized by diffuse fibrous thickening of the leaflets and fusion of two or three commissures. Leaflet thickening usually occurs in the absence of calcific deposits, and the anteroseptal commissure is most commonly involved. Incompletely developed leaflets, shortened or malformed chordae, a small annulus, or an abnormal number or size of papillary muscles may result in Congenital TS. Genetic or acquired/environmental causes can disrupt the normal organization and composition of the extracellular matrix as well as communication between valve endothelial cells and interstitial valve cells. Finally, it alters valve mechanics and interfere with the valve leaflet function, culminating in heart failure. [6] The primary result of TS is right atrial pressure elevation and consequent right-sided congestion.

CASE SUMMARY

46 years female, farmer by occupation and residing in a remote village of Nepal presented with NYHA-III shortness of breath and swelling of the body for 6 months. She has past history of Percutaneous Transluminal Mitral Commissurotomy (PTMC) done in 2013 AD for symptomatic severe rheumatic mitral stenosis. She felt better symptomatically for few years but again became symptomatic.

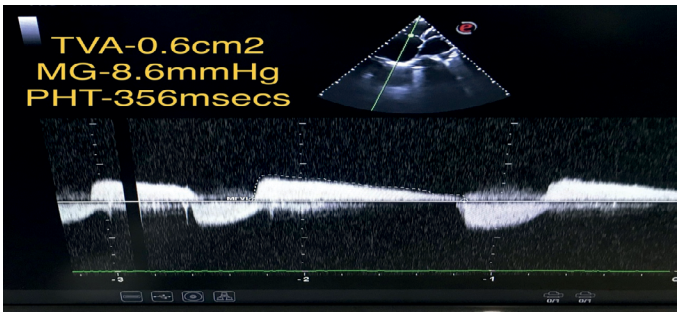

Figure 1: Echocardiography shows severe TS with Tricuspid Valve Area-0.6cm2, Mean Gradient-8.6mmHg and Pressure Half Time-356msecs.

She has additionally Type 2 Diabetes Mellitus (DM) with hypothyroidism with anemia as additional comorbidities. Clinical examination revealed irregular pulse with pulse deficit of 20 bpm, bilateral pitting pedal edema, raised Jugular Venous Pressure (JVP), tapping apex, Left parasternal heave, Apical mid-diastolic thrill, Left lower border Mid-diastolic murmur, Pansystolic murmur over tricuspid area and inspiratory basal crackles over chest.

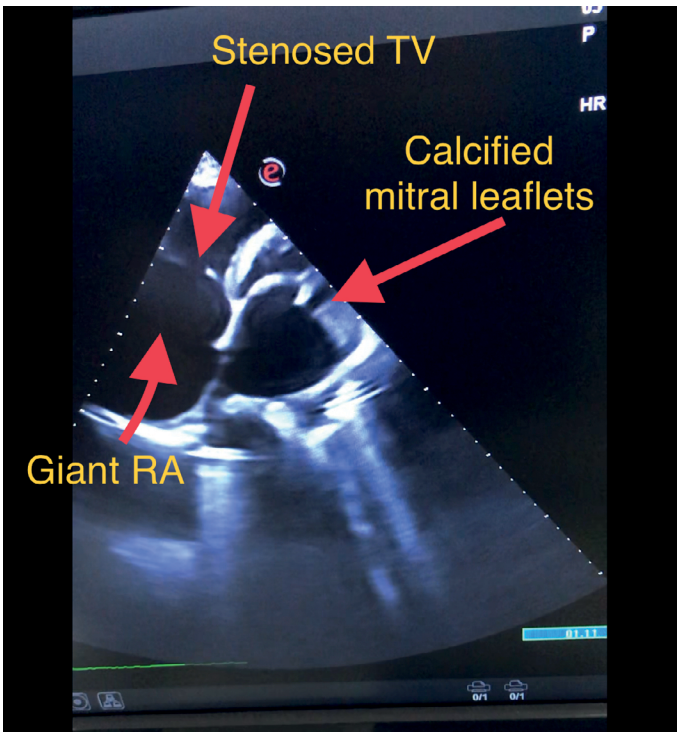

Figure 2. Echocardiography shows Severe Mitral Stenosis with calcified mitral leaflets

Echocardiography evaluation (shown in Fig 1 and 2) suggests re-stenosed mitral valve (Mitral Valve Area-1.6cm^2 by planimetry) with calcified mitral leaflets, Dilated Left Atrium, Mild Aortic Regurgitation, Severe TS (Mean Gradient-8.6mmHg, Pressure Half Time-356msecs, Tricuspid valve area 0.6cm^2), Severe TR, Mild Pulmonary Arterial Hypertension (Pulmonary Artery Systolic Pressure-32mmHg), Giant Right Atrium, Dilated Right Ventricle, Inferior Venacava and Hepatic Veins.

Electrocardiogram (shown in Figure 3) reflects fibrillatory P waves with irregularly irregular rhythm suggestive of Atrial fibrillation (AF) with controlled ventricular rate.

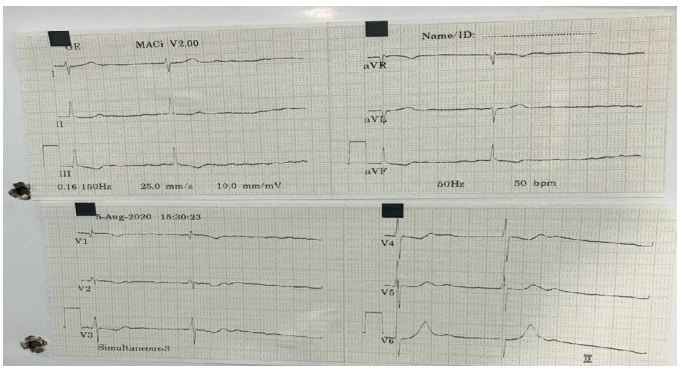

Figure 3. Electrocardiogram(ECG) shows Atrial Fibrillation

She has been managed with medical therapy including diuretics, oral penicillin, warfarin and digoxin. Also, she has been treated with metformin and thyroxine for DM and hypothyroidism. She has been advised for surgical treatment including mitral commissurotomy with Tricuspid valve repair or replacement.

DISCUSSION

Most stenotic tricuspid valves are associated with clinical evidence of regurgitation that can be documented by performing physical examination (murmur) and Echocardiography. The low cardiac output characteristic of TS causes fatigue, and patients often experience discomfort caused by hepatomegaly, ascites, and anasarca. Despite the coexistence of Mitral Stenosis (MS), the symptoms characteristic of this valvular lesion (severe dyspnea, orthopnea, and paroxysmal nocturnal dyspnea) are usually mild or absent in the presence of severe TS because the latter prevents surges of blood into the pulmonary circulation behind the stenotic mitral valve.

Mean gradient greater than or equal to 5 mm Hg at normal heart rate is considered indicative of clinically significant TS. Higher gradients may be seen with combined stenosis and regurgitation. A longer T 1/2 (pressure halftime by continuous wave Doppler) implies a greater TS severity with values of greater than or equal

to 190 ms being frequently associated with significant (or critical) stenosis. Other parameters that are seen include-reduced Tricuspid Annular Plane Systolic Excursion (TAPSE), dilated IVC, and increased right atrial size. [7]

Although the fundamental approach to the management of severe TS is surgical treatment, intensive sodium restriction and diuretic therapy may diminish those symptoms secondary to the accumulation of excess salt and water. Treatment of SLE and APLA syndrome may reduce the "coating" over the valves and chordae and reduce stenosis and regurgitation.[8] Cessation of fenfluramine or methysergide has been associated with valve normalization in drug induced stenosis.[9]

Most patients with TS have coexisting valvular disease that requires surgery. In patients with combined TS and MS, the former must not be corrected alone because pulmonary congestion or edema may ensue. Surgical treatment of TS should be carried out at the time of mitral valve repair or replacement in patients with TS in whom the mean diastolic pressure gradient exceeds 5 mm Hg and the tricuspid orifice is less than approximately 2.0 cm2. The final decision concerning surgical treatment often is made at the operating table. [3]

Closed Valvotomy is performed using 1, 2, or 3 size balloons. Yash Y et al.[10] reported a patient with congenital tricuspid stenosis successfully treated by percutaneous balloon valvotomy that appears to be an alternative to surgery. Open valvotomy in which the stenotic tricuspid valve is converted into a functionally bicuspid valve may result in substantial improvement. The commissures between the anterior and septal leaflets and between the posterior and septal leaflets are opened.

If open valvotomy does not restore reasonably normal valve function, the tricuspid valve may have to be replaced. A large bioprosthesis is preferred to a mechanical prosthesis in the tricuspid position because of the high risk of thrombosis of the latter and the longer durability of bioprostheses in the tricuspid than in the mitral or aortic positions. [3] However, in carcinoid syndrome, a mechanical valve is preferred over bioprosthetic to avoid degeneration.

Surgery for severe TS and TR is most often performed at the time of operation for left-sided valve disease, chiefly rheumatic mitral stenosis/mitral regurgitation. Tricuspid valve surgery is preferred over percutaneous balloon tricuspid commissurotomy for treatment of symptomatic severe TS and TR. In our patient, we have referred for possible Bioprosthetic Tricuspid valve replacement and Mitral valve repair.

CONCLUSION

Rheumatic Heart Disease may progressively present with additional combined rare valvular lesions like severe tricuspid stenosis. The prognosis of patients diagnosed with Severe TS is bleak with many requiring valve surgeries. So, RHD patients should be on regular follow up and assessed periodically with Echocardiography to avoid dreaded life-threatening complications.

REFERENCES

1. Tao G, Kotick JD, Lincoln J. Heart valve development, maintenance, and disease: the role of endothelial cells. Curr Top Dev Biol. 2012;100:203-32. [DOI]
2. Waller BF. Morphological aspects of valvular heart disease: Part I. Curr Probl Cardiol. 1984;9(8):1-66. [DOI]
3. Catherine M. Otto and Robert O. Bonow.Valvular heart disease. In: Douglas L. Mann, Douglas P. Zipes, Peter Libby, Robert O. Bonow, Eugene Braunwald., editors. Braunwald's Heart Disease: a Textbook of Cardiovascular Medicine. Philadelphia, PA:Elsevier/ Saunders, 2020;1497-99.
4. Sharieff S, Saghir T, Shah-e-Zaman K, et al. Concurrent percutaneous valvuloplasty of mitral and tricuspid valve stenoses. J Invasive Cardiol 2005;17(6):340-2. [Google Scholar]
5. S. Laudari and G. Subramanyam. A study of spectrum of rheumatic heart disease in a tertiary care hospital in Central Nepal. Int J Cardiol Heart Vasc. 2017; 15: 26-30. [DOI]
6. Hinton RB Jr, Lincoln J, Deutsch GH, Osinska H, Manning PB, Benson DW, Yutzey KE. Extracellular matrix remodeling and organization in developing and diseased aortic valves. Circ Res. 2006 Jun 9;98(11):1431-8 [DOI]
7. Baumgartner H, Hung J, Bermejo J, Chambers JB, Evangelista A, Griffin BP, et al.; American Society of Echocardiography; European Association of Echocardiography. Echocardiographic assessment of valve stenosis: EAE/ASE recommendations for clinical practice. J Am Soc Echocardiogr. 2009 Jan;22(1):1-23; quiz 101-2 [DOI]
8. Adler DS. Non-functional tricuspid valve disease. Ann Cardiothorac Surg. 2017 May;6(3):204-213 [DOI]
9. Seghatol FF, Rigolin VH. Appetite suppressants and valvular heart disease. Curr Opin Cardiol. 2002 Sep;17(5):486-92. [DOI]
10. Yash Y. Lokhandwala, Rajesh M. Rajani, Bharat V. Dalvi and Purshottam A. Successful balloon valvotomy in isolated congenital tricuspid stenosis. CardioVascular and Interventional Radiology, Springer New York 1990;13(6):354-56. [DOI]