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A Rare Case of Isolated Rheumatismal Tricuspid Valve Involvement in a Middle-Aged Woman

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ABSTRACT

Introduction: Tricuspid valve stenosis (TS) is often combined with tricuspid regurgitation and most cases of TS have a rheumatic origin. TS is usually associated with other valvopathies when occurring in a rheumatic context. Rheumatismal tricuspid stenosis typically occurs with concomitant mitral and/or aortic involvement. Other causes are rare, including congenital, carcinoid, and drug-induced valve disease, Whipple`s disease, endocarditis, and large right atrial tumor.
Case Presentation: Here, we present a 42-year-old symptomatic woman with isolated TS due to rheumatic heart disease.
Conclusion: Patients suffering from rheumatic heart disease should be on regular follow-ups and periodically assessed with echocardiography. In the present case, the tricuspid valve was involved in isolation.

1. Introduction

In clinical practice, native tricuspid stenosis (TS) is rare (1). A common etiology of TS is iatrogenic, developing after repairs or replacements of the tricuspid valve (TV) (2). Other causes of native TS include rheumatic disease, genetic abnormalities, metabolic or enzymatic abnormalities (e.g., carcinoid disease, Fabry's disease, Whipple's disease, methysergide treatment), and active infective endocarditis (3).

Rheumatic heart disease (RHD) occurs following rheumatic fever caused by group A streptococci (4). In the case of patients with RHD, TS is present only in 15% of autopsies and approximately 5% of symptomatic cases (5). In most cases, rheumatic TS is accompanied by concomitant left-sided valvular involvement. The first isolated TS of rheumatic origin was described by Morgan *et al.* in 1971 (6).

Diffuse fibrous thickening of the leaflets and fusion of two or three commissures distinguish rheumatic tricuspid valve disease (7). In the absence of calcific deposits, leaflet thickening mostly occurs at the anteroseptal commissure. Treatment is usually unnecessary for isolated mild TS, but surgical repair or replacement is necessary if other heart valves are damaged (8). Here, We describe a patient with isolated rheumatic TS.

2. Case Presentation

A 42–year-old housewife presented with fatigue, generalized edema, and New York Heart Association (NYHA) functional class-II shortness of breath and was admitted to the hospital. She remembered that her symptoms started at the age of twenty. When she was a child, she experienced migratory polyarthritis. She had no history of scleroderma, systemic lupus erythematosus (SLE), drug ingestion, radiation exposure, additional comorbidities, or past cardiac interventions.

On examination, she had generalized edema. Her jugular vein was engorged. Her pulse rate was irregular, with 80 beats per minute. A diastolic murmur was heard along the left parasternal border in the fourth intercostal space, and a systolic murmur was prominent at the apex. Echocardiography evaluation revealed thickened, highly calcified, and tethered tricuspid valve leaflets with a large coaptation defect resulting in severe tricuspid regurgitation (Figure 1) and severe TS with an average mean gradient of 11 mmHg (Figure 2) and pressure half-time (PHT) of 222 msec, but no echocardiographic signs of Ebstein anomaly.

The result of 3D planimetry showed that the tricuspid valve had a commissural fusion with chordal thickening.

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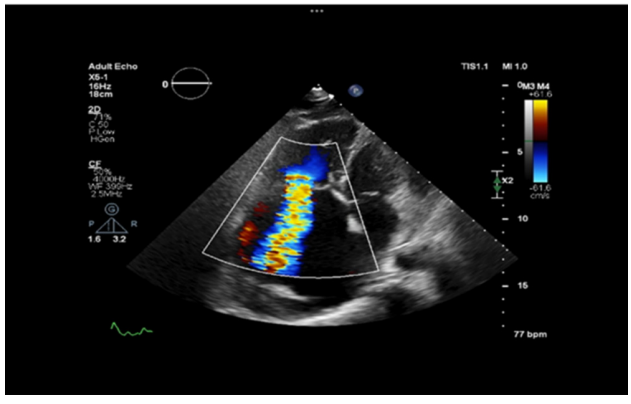

Figure 1. Transthoracic Echocardiography Image Showing Severe Tricuspid Stenosis with Severe Tricuspid Regurgitation.

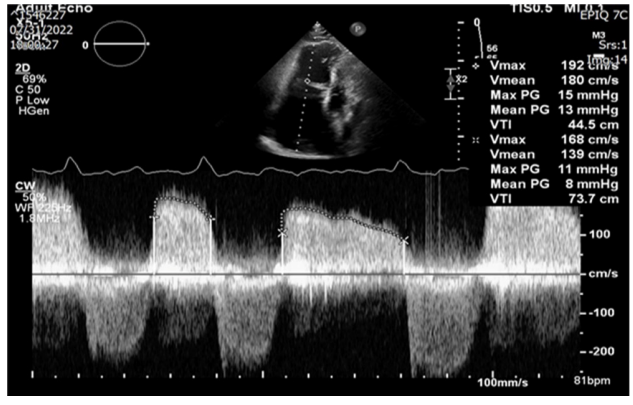

Figure 2. Transthoracic Echocardiography Showed Severe Tricuspid Stenosis with Mean Gradient = 11 mmHg and Pressure Half Time = 222 msec.

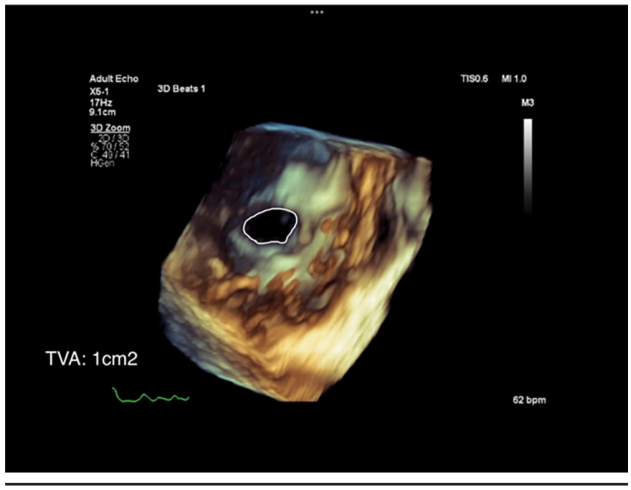

Figure 3. Three-Dimensional Echocardiography Image Revealing Severe Tricuspid Stenosis with Tricuspid Valve Area (TVA) = 1 cm^2.

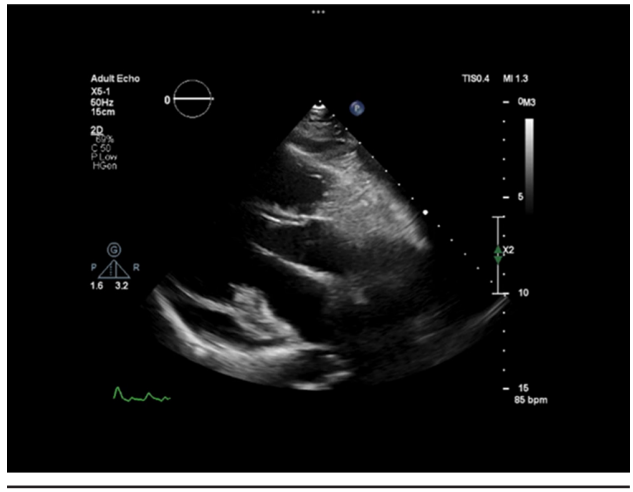

Figure 4. Transthoracic Echocardiography Showed no Rheumatismal Changes in the Mitral Leaflets.

The tricuspid valve area was reduced to 1 cm^2 (Figure 3) due to rheumatic involvement, associated with a large systolic coaptation defect, resulting in torrential tricuspid regurgitation; the tricuspid regurgitation gradient (TRG) was estimated at 26 mmHg. The mitral valve leaflets were seen without rheumatismal change; the posterior leaflet was hypoplastic with mild to moderate mitral regurgitation but no mitral stenosis (Figure 4).

The aortic and pulmonic valves were also normal (Figure 5). The left ventricular volume was reduced. The right atrium and the right ventricle were enlarged due to severe tricuspid regurgitation. The inferior vena cava (IVC) and hepatic vein were dilated. The pulmonary artery systolic pressure was 41 mmHg, interpreted as mild pulmonary hypertension.

We prescribed furosemide 20 mg three times a day, spironolactone 12.5 mg daily, bisoprolol 2.5 mg daily, and rivaroxaban 5 mg daily, besides an intramuscular injection of penicillin 1,200,000 IU every four weeks. Her symptoms improved for a month but then worsened. Hence, a tricuspid valve surgery was recommended to the patient, which she refused.

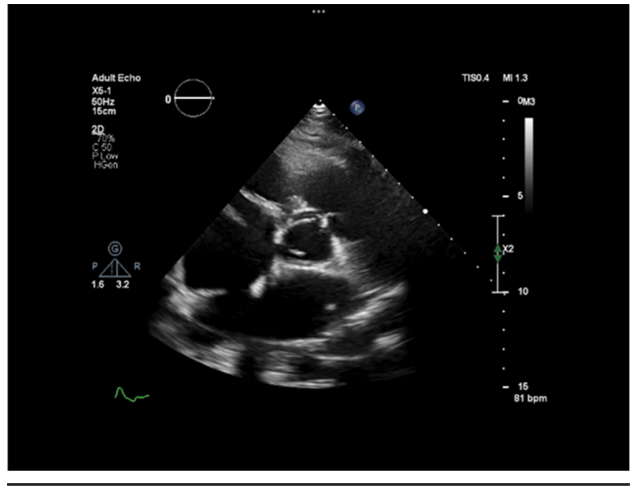

Figure 5. Transthoracic Echocardiography Image Showing Normal Aortic and Pulmonary Valves.

3. Discussion

Most tricuspid stenosis (TS) cases are concomitant with evidence of clinical regurgitation (9, 10). TS is usually rheumatic in origin and accompanied by mitral and aortic valve involvement (10). Severe TS and high right atrium pressure lead to venous congestion, hepatomegaly, ascites, and anasarca, resulting in fatigue and discomfort (11). In the absence of genetic reasons or active infective endocarditis, the development of TS takes many years (12). Despite the concomitance of mitral stenosis with severe TS in many cases, the symptoms are usually mild or absent because the latter prevents blood surges into the pulmonary circulation

behind the stenotic mitral valve (13).

Tricuspid stenosis is considered severe when the mean gradient is ≥ 5 mmHg at a regular heart rate (3). A higher gradient can be observed when stenosis is combined with regurgitation. A PHT ≥ 190 msec also implies severe TS when assessed by continuous wave Doppler (3). Other parameters include tricuspid annular plane systolic excursion (TAPSE) decrement, dilated IVC, and right atrial size enhancement.

The essential approach to severe TS management is a surgical procedure. However, severe sodium intake restriction and diuretic therapy could reduce symptoms (14). The treatment of SLE and antiphospholipid antibodies (APLA) syndrome may decrease the coating over the valves and chordae, thereby reducing stenosis and regurgitation (15, 16). The termination of fenfluramine or methysergide is related to valvular normalization in drug-caused stenosis (17). Surgical treatment of TS should be performed during mitral valve repair or replacement. Taken together, tricuspid valve surgery is preferred over percutaneous balloon commissurotomy to treat severe symptomatic TS and tricuspid regurgitation (18). In the present case, tricuspid valve surgery was recommended to the patient.

Conclusion

Rheumatic heart disease (RHD) may progressively present with additional combined rare valvular damage like severe TS. Patients suffering from RHD should be on regular follow-ups and periodically assessed with echocardiography. In the present case, an isolated tricuspid valve was involved due to rheumatic fever.

Informed Consent

Written informed consent was obtained from the patient to publish this case report.

Ethical Approval

This study was approved by the institutional ethical committee under the ethical approval code of IR.RHC.REC.1402.031.

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Authors' Contribution

Study concept and design: F.K, collection, and interpretation of data: S.M and F.A, drafting of the manuscript: M.S.A and L.A and P.A, revising the manuscript: A.S.

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Financial Disclosure

The authors declare no conflicts of interest.

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