Isolated Tricuspid Stenosis in a Patient with History of Scleroderma

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Abstract

Isolated tricuspid stenosis (TS) is a very uncommon finding. There are some reports of isolated TS in the setting of systemic lupus erythematous, but its presence in scleroderma has never been reported before. We report a 48-year- old female with isolated TS with a background of scleroderma, diagnosed by echocardiography ***(Iranian Heart Journal 2012; 13 (1):52 -54*).**

Case report

The patient was a 48-year-old female with a previous diagnosis of scleroderma made 16 years previously based on Reynaud phenomenon, telangectasia, and limited skin involvement . She also suffered from systolic hypertension since the initial diagnosis was made.

She was referred to our center with complaints of newly found dyspnea on exertion of a 6-month duration, which exacerbated from NYHA function class 1 to 3 in this timeline. On presentation, she was normotensive with BP of 125/75 mmHg and found to have signs and symptoms suggestive of right-sided heart failure, which included increased JV pressure, mild ascites, and lower extremity edema. On cardiac auscultation, she had 3/6 mid-systolic murmur best heard on the right sternal border, which showed increase in intensity with inspiration and was mostly consistent with TS. Other aspects of the cardiac physical examination were unremarkable.

There were no positive findings in favor of endocarditis. Electrocardiographic findings pointed to LV hypertrophy voltage criteria and RA abnormality and there were no conduction defects.

Her medications included Captopril 50 mg BID and she did not use any other drugs at the time.

In her laboratory studies, her ESR level was in normal range , CRP was 1+ positive , and other lab tests including serum Cr were in normal range. Blood cultures were consistently negative.

On echocardiographic evaluation, we found a normal left ventricular size and preserved LV function , normal RV size and moderate systolic dysfunction , no left valvular disease , no pulmonary valve disease , RA enlargement with RA area= 30 cm2 , moderate pericardial effusion , thickened TV with severe TS (mean gradient = 14 mmHg , peak gradient = 23 mmHg) , mild TR, and no typical Ebstein anomaly(distance between MV and TV = 7.8 mm/m2) (Figs. – 1, 2, 3).

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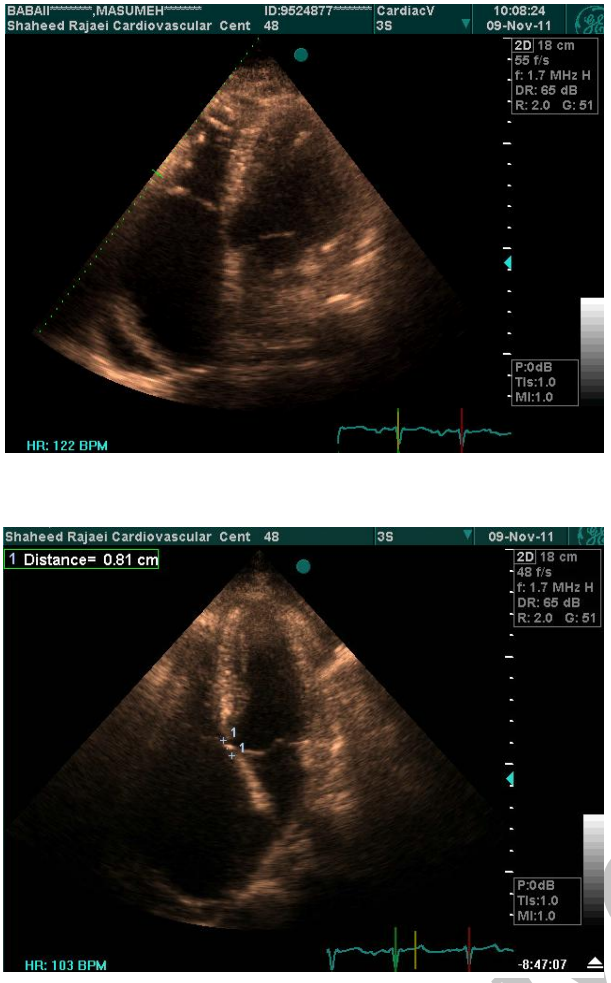

Fig. 2. Thickened TV .Pericardial effusion around RA Figure 2 -Non typical Ebstein anomaly (distance between MV and TV = 7.8 mm/m2)

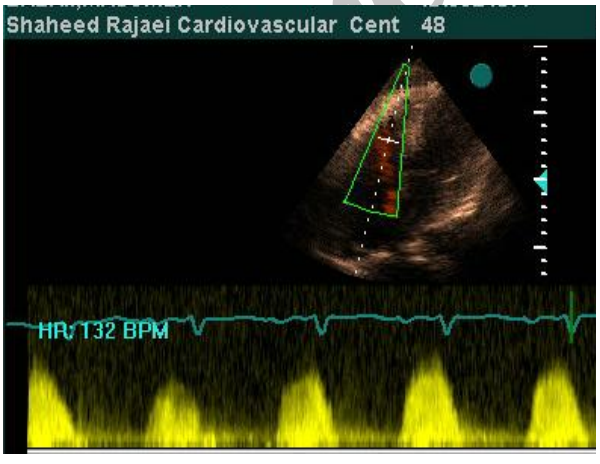

Fig. 3. Severe TS (mean gradient = 14 mmHg , peak gradient = 23 mmHg)

Discussion

Usual cases of tricuspid valve stenosis encountered in practice are mostly in the setting of rheumatic heart disease and are usually accompanied by other valvular lesions, and isolated involvement is extremely rare. TS in RHD is mostly like mitral stenosis with fusion in the chordate and thickening of the leaflets. For the diagnosis of TS to be made, we have to demonstrate that the diastolic gradient between the right atrium and right ventricle is significantly elevated even at 2 to 5 mmHg mean pressure. Accordingly, right atrial pressure increases and this will lead to venous congestion and distention of the jugular veins, ascites, pleural effusion, and peripheral edema.[1] The right atrial wall thickens, and the atrial chamber dilates if the patient remains in normal sinus rhythm. Additionally, the right atrial tracing and jugular venous pulse will show prominent a waves and accentuation with inspiration. The cardiac murmur is mid-diastolic, increases with inspiration, is heard maximally along the left sternal border, and may have an opening snap.

Other causes beside RHD include carcinoid syndrome, SLE, right atrial myxoma, congenital atresia, and infiltrating tumors.[2] There have never been reports of isolated TS in scleroderma. Cardiac involvement in scleroderma is mostly seen in the form of pericardial involvement, necropsy on myocardial biopsy specimen, vasospastic incidents and conduction defects. Also, hypertensive heart disease is common but valvular involvement is not one of its usual features.[4]

Valvular involvement in SLE is a known entity and there are some reports of isolated TS in the setting of this autoimmune disease. It is very important to rule out infectious endocarditis because missing this would have catastrophic effects. Giving common overlap syndromes in rheumatic heart diseases and relatively unknown pathophysiological basis for valvular stenosis in their setting, it is

possible that the underlying mechanism in this entity is shared between these diseases.[5] Until further evaluation and studies management strategies and options, it is advisable that the recommended guidelines for rheumatismal TS be adhered to.

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