CASE REPORT

Isolated Tricuspid Valve Stenosis - Probably of Rheumatic Etiology

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Abstract

Rheumatic heart disease is still a common cardiac disease in developing countries. Tricuspid valve is not usually involved in RHD and whenever it is involved, it is always associated with mitral valve disease. We report a case of isolated tricuspid stenosis, most likely of rheumatic etiology.

Case

A 25 years female presented to us with history of periorbital puffiness for 2 years. Initially the puffiness used to appear in morning, but later on, it was present throughout the day, more so on rising from bed in morning. Patient also complained of easy fatigability for the last two years which was gradually progressive. She complained of pedal edema for the last two weeks. There was no H/O fever, joint pains or dyspnoea. There was no H/O decreased urinary output or hematuria. With these symptoms she went to the local hospital, where she was investigated for renal disease, but her renal function test including ultrasound of abdomen was reported to be normal. Then she was referred to us.

On examination, her blood pressure was 100/80 mmHg, pulse 76/min. Her JVP was raised with prominent a wave. Her apex beat was in 5th intercostals space, medial to mid clavicular line. First heat sound was loud, 2nd heart sound was normal. There was mid diastolic murmur, medial to apex, with presystolic accentuation, which increased on inspiration. There was no other murmur. Liver was enlarged 5cm below costal margin with diastolic pulsations. Keeping in view, the prominent a wave in JVP, the long mid diastolic murmur, absence of evidence of PAH and no dyspnea, we made the diagnosis of tricuspid valve stenosis (TS) with probably mitral stenosis (MS), as isolated TS is almost unknown.

Investigations

- Hb 12gm /dl
- TLC: 8500/cubic mm
- DLC: polys 59, lymphocytes 34, monocytes 4, eisonophylls 2
- Urea 27mg%
- Creatinine 1.0 mg%
- Fasting blood sugar 76mg%
- Serum electrolytes
  - sodium 148meq/l
  - potassium 4.7meq/l
  - chloride 104meq/l
- Bilirubin 3mg/dl

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Conjugated: 1mg/dl
Platelet count 1.9 lac/cmm
5 hydroxyindoleacetic acid(5-HIAA) in urine: 1.19mg/g creatinine(normal<10.00)
Anti phospholipids antibody, IgM: 2.54 MPL/ml (normal<10.00)
Lupus anticoagulant: absent
ECG showed right atrial enlargement
X ray chest showed, cardiomegaly with right atrial enlargement and prominent superior vena caval shadow.
Echocardiography showed normal mitral, aortic and pulmonary valves. Tricuspid valve(TV) was thickened and doming, with thickened and fused chordae (Figure 1).Continuous Doppler examination across TV showed significant gradient (Figure 2) Peak gradient was 15 mm Hg and mean gradient was 10.2 mmHg.

So the final diagnosis was isolated tricuspid valve stenosis. Etiology could be rheumatic or congenital, but more likely it was of rheumatic.

**Discussion**

TS is a rare disorder. Around 15 % patients with rheumatic heart disease at autopsy show evidence of TS. But it is clinically significant in less than 5% of cases.

TS is almost always rheumatic in origin. It is almost always associated with mitral and aortic valve involvement. TV shows diffuse thickening of leaflets with or without fusion of commissures. Chordae tendineae are usually thickened and shortened. Deformity of the valve tissue commonly leads TS with TR. It occurs more commonly in females. Congenital form of disease is very rare and its true incidence is not known. It is more commonly seen in infants. Leaflets are usually incompletely developed, chordae are shortened and malformed, small annuli. There may be abnormal number and size of papillary muscles or there may be a combination of these defects.

Other rare causes of TS include right atrial tumor, carcinoid syndrome, vegetation on the tricuspid valve, and pacemaker lead. Carcinoid valve lesions characteristically manifest as fibrous white plaques located on the valvular and mural endocardium. The valve leaflets are thickened, rigid, and reduced in area. Fibrous tissue proliferation is present on the atrial and ventricular surfaces of the valve structure. Large infected vegetations obstructing the orifice of the tricuspid valve may produce stenosis. This condition is relatively uncommon, even in those who abuse intravenous drugs. Mukhopadhyay et al reported a case producing features of TS because of primary antiphospholipid syndrome, presenting as intracardiac thrombus over the tricuspid valve.

Our case was having isolated stenosis of TV. There was no associated TR. There was no evidence of carcinoid, infective endocarditis, thrombus. There was no history of rheumatic fever in the past. Mitral valve was normal.

Since the congenital TS usually presents in infancy,
annulus is small, leaflets are usually incompletely developed, which was not seen in echocardiography in the present case. So in all probability, it was a case of isolated TS of rheumatic etiology. The patient presented at 25 years of age, Echocardiographic picture of tricuspid valve was same as we see in rheumatic TS with associated mitral valve disease. History of rheumatic fever may not be present in around 50% patients of RHD. There are few case reports of isolated TS of rheumatic etiology in the literature.\(^4,5\)

Medical management of TS includes sodium restriction and diuretic therapy. However, the disease is progressive and surgical therapy is often required. The operative treatment of TS has involved closed commissurotomy, open commissurotomy, and open valvuloplasty (balloon valvuloplasty has also been tried but with varying degrees of success).

**References**