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Severe Isolated Rheumatic Aortic Valve Regurgitation

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Dear Editor,

Aortic valve (AV) disease constitutes an important cause of morbidity and mortality worldwide. Among the etiologies of AV disease, rheumatic involvement remains prevalent in the developing world.

In our echo ward, we have encountered 3 similar cases in recent months. Herein, we describe one of them: a 30-year-old woman presenting with dyspnea (functional class II-III) and palpitation of 2 years' duration. Her symptoms exacerbated during sleep. On physical examination, she had no stigmata of Marfan's syndrome or any sign or symptom of rheumatologic disease. Her blood pressure and heart rate were normal with a wide pulse pressure (bounding pulses), a prominent point of maximal impulse, a systolic murmur (II/VI) in the aortic area, and a holodiastolic murmur (IV/VI) at the left sternal border. ECG showed a normal sinus rhythm with an inverted T wave in leads V_1 and V_2. Laboratory tests were normal. Chest X-ray revealed cardiomegaly without any other abnormalities. Transthoracic and transesophageal echocardiographic examinations showed a mild increase in left ventricular (LV) volume (end-diastolic volume = 79 mL/m^2) and a mildly reduced ejection fraction (50%). In addition, there were a tricuspid AV and irregular thickening of the leaflets (especially their tips), retracted cusps, and a central triangular coaptation defect resulting in a severely eccentric aortic regurgitation (AI) jet with a holodiastolic flow reversal in the descending thoracic and abdominal aortas. The other valves were completely normal, and so was the size of the ascending aorta.

The patient was scheduled for AV replacement due to severe symptomatic AI.

In this case, we had an isolated involvement of the AV in a young woman without any past medical history.

There are 2 major reasons for AI:

1) Conditions affecting the valve

2) Conditions affecting the aorta and only secondarily causing valvular incompetence (1)

Rheumatic heart disease continues to be the dominant form of heart valve disease in developing nations (2). Anatomically, AV disease causing stenosis or regurgitation or both has been considered by numerous investigators generally to be of a rheumatic etiology, albeit concomitant with mitral valve involvement (3). Of course, rheumatic heart disease was the most common cause of isolated AI in the older surgical reports of Mayo Clinic, whereas in the more recent series, aortic dilation and/or degenerative valve change comprise half the cases (4). Accordingly, in patients with a normally functioning mitral valve, rheumatic heart disease is an infrequent cause of pure AI.

All the following morphological features of the rheumatic heart disease of the AV (mentioned in previous literature for individuals aged < 35 y) and all the following parameters of severe AI were present in our patient:

Rheumatic involvement features:

- Irregular or focal thickening
- Coaptation defect
- Restricted leaflet motion

And for severe AI:

- Confirmed severity in 2 echocardiographic views
- Jet lengths $\geq$ 1cm in at least 1 view
- Velocity $\geq$ 3m/s in early diastole
- Pandiastolic jet in at least 1 envelope
- Holodiastolic flow reversal in the descending and abdominal aortas (5)

Consequently, in our patient, irrespective of the absence of the involvement of the other valves, all echocardiographic criteria showed that the etiology of her pure severe AI must have been rheumatic.

Finally, our patient underwent surgery and pathological examination confirmed the severely rheumatismal in-

volvement of the AV with fibrosis and scaring. It is deserving of note that our other 2 patients also had isolated severe AI.

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