A Severe Restrictive Aortic Regurgitation Resulting From Valve Tenting by Unusual Aortic Chordae Tendineae Strands

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We report a case of a 74-year-old man who presented with progressive dyspnea and hypertension (200/63 mm Hg) without any chest pain. He had a history of chronic hypertension and heart murmur several years ago. Physical examination revealed a diastolic murmur with an acute pulmonary edema that required admission in the intensive care unit. The ECG results were normal. There was no fever and no inflammatory syndrome; hemocultures were sterile. Transthoracic echocardiography (Figure 1, online-only Data Supplement Movies I and II) showed a severe aortic regurgitation on a mildly thickened valve with a mild dilatation of the aortic root (Valsalva aortic diameter = 46 mm; 25 mm/m²) with a normal annulus diameter (22 mm) and suspected intra-aortic mobile linear echoes.

Transesophageal echocardiography (Figure 2, online-only Data Supplement Movies III through VI) showed a tricuspid aortic valve with a large coaptation defect resulting in a massive aortic regurgitation with a mild dilatation of the aortic root and a normal annulus. Several linear mobile echoes looking like chordae tendineae strands were connecting the Valsalva aortic root to the sigmoid cusps, without any...

Figure 1. Transthoracic echocardiography (parasternal longitudinal view) showing severe aortic regurgitation with mild dilatation of the aortic root (Valsalva 46 mm), a normal annular diameter (22 mm), and suspected intra-aortic mobile linear echoes (arrows).
dissection flap or chordae rupture. The aortic valve was moderately thickened with a normal opening movement, but with an important tenting of the cusps that could not close normally (with the anterior leaflet presenting a plicature), resulting in a severe restrictive aortic regurgitation. The left ventricle was slightly dilated and hypertrophied with a preserved ejection fraction (55%), and there was a mild mitral regurgitation.

Because of this symptomatic severe aortic regurgitation, the patient underwent a surgical replacement of the aortic valve, which was tricuspid, thickened, and fenestrated with several chordae tendineae strands suspending the cusps to a dilated aortic root with a normal annular diameter and a thin aortic wall (Figure 3A through 3C). The surgeon made a Bentall intervention by replacing the aortic valve with a biological prosthesis and the aortic root by a prosthetic graft. The patient was well 2 years after surgery.

Chordae tendineae strands have rarely been described on bicuspid and tricuspid aortic valves, with aortic regurgitation due to spontaneous rupture of chordae.1–3 One case reported a severe aortic regurgitation resulting from a congenital downward displacement of anterior aortic annulus on a bicuspid valve with fibrous strands.4 To our knowledge, our case is the first to describe this mechanism of aortic regurgitation due to the tenting of a tricuspid aortic valve by chordae attached to a dilated aortic root, without annular dilatation or chordae rupture, and without any congenital heart defect (Figure 4). The chronic hypertension probably played a role in the aortic dilatation, and aging, as well. The aortic chordae tendineae strands may be embryonic remnants of the cusp formation process at an early stage of the aortic valve development, which could leave fibrous tissue between the aortic valve and the aortic wall.5 However, to our knowledge, embryological studies do not yet support this.6

Figure 2. Transesophageal echocardiography showing the chordae tendineae strands (arrows) connecting a mildly dilated aortic root to sigmoid cusps that present tenting and restriction with a severe aortic regurgitation.

Figure 3. A and B. Surgical views of the aortic valve with several chordae tendineae strands (arrows) suspending the cusps to a mildly dilated aortic root. C, Isolated aortic valve with chordae tendineae strands.
not mention aortic chordae remnants. Such chordae tendineae strands may be difficult to see at transthoracic and even transesophageal examinations, and they might be more frequent than previously known. They must be carefully searched, in particular, when echography shows linear mobile echoes on the aortic valve or an unexplained aortic regurgitation.

Disclosures

None.

References

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