A Case of Severe Aortic Regurgitation Caused by Takayasu’s Arteritis Showing End-Diastolic Opening of Aortic Valve

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INTRODUCTION

The hemodynamic manifestations of acute severe aortic regurgitation (AR) are characterized by a steep increase in left ventricular (LV) end-diastolic pressure (EDP), exceeding mean left atrial pressure and occasionally equilibrating with aortic diastolic pressure. Diastolic opening of the aortic valve (AV) in severe AR is an exceptionally rare phenomenon, which could be caused by equalization of LV EDP and systemic blood pressure due to severely elevated LV EDP.1-9

We report a case of Takayasu’s arteritis showing diastolic opening of the AV due to severe AR associated with aortic root dilatation, in which unique hemodynamics were successfully evaluated using various echocardiographic findings.

CASE PRESENTATION

A 42-year-old man was transferred from another hospital for worsening heart failure as a result of AR. Two months prior to this transfer, he came to the hospital because of a 9-month history of dyspnea on exertion and was diagnosed with acute decompensated heart failure on the basis of the finding of lung congestion. He was admitted to the hospital and diuretic therapy was initiated, but complete relief of dyspnea was not achieved, and severe AR was detected. Upon admission after the transfer, he denied dyspnea at rest, and physical examination revealed no jugular venous distention, peripheral edema, or rales in the lung fields. The body temperature was 36.1°C. His blood pressure was 104/44 mm Hg, and pulsus magnus and pulsus celer were observed, suggesting the presence of hemodynamically significant AR. A systolic ejection murmur and diastolic murmur were heard maximally at the second left sternal border. Electrocardiography revealed sinus rhythm with signs of left atrial enlargement and LV hypertrophy. Chest radiography displayed slight cardiomegaly without lung congestion. Blood examination showed decreased hemoglobin concentration (107 g/L) and markedly elevated C-reactive protein (9,300 μg/L) and plasma brain natriuretic protein (118.2 pg/mL).

Transthoracic echocardiography was performed to evaluate AR. Two-dimensional images showed that the left ventricle was enlarged with eccentric hypertrophy (end-diastolic dimension 61 mm, relative wall thickness 0.26, indexed LV mass 121 g/m²), whereas LV ejection fraction was preserved at 66% (Videos 1 and 2). Although LV ejection fraction was preserved, mitral annular peak systolic velocity and LV global longitudinal strain were reduced (7 cm/sec and −14%, respectively), suggesting intrinsic myocardial damage. Color Doppler images showed severe AR (Figure 1, Video 3), with vena contracta width of 11 mm. The severity of AR was further confirmed by a regurgitant volume of 70 mL quantified using the proximal isovelocity surface area method and obvious holodiastolic flow reversal of abdominal aortic blood flow (Figure 2). The AV did not show any organic abnormality, but coaptation was limited by the enlarged aortic root. The anterior-to-posterior diameters of the aortic annulus, sinus of Valsalva, sinotubular junction, and ascending aorta were 22, 43, 41, and 42 mm, respectively (Figure 3). Therefore, the cause of AR was diagnosed as tethering due to aortic root dilatation (El Khoury type Ib). Surprisingly, the AV showed late diastolic opening, occurring just after the P wave of the electrocardiogram (Figure 4, Video 4). The late diastolic opening of the AV was suspected to be the result of marked elevation of LV EDP. Consistent with this hypothesis, transmitral Doppler flow showed a lack of late diastolic inflow along with prominent atrial systolic backward flow in the pulmonary venous Doppler flow (Figure 5), suggesting markedly elevated LV EDP. Late diastolic mitral regurgitation was observed (Video 5), which also suggested elevated LV EDP. Furthermore, the continuous-wave Doppler waveform of the AR showed extremely shortened pressure half-time (174 msec) and abrupt decline of velocity just before atrial contraction to end-diastole (Figure 6A), and precise observation of the color Doppler image as well as color M-mode Doppler image of the AV revealed discontinuation of regurgitation at late diastole (Figure 6B, Video 3), indicating equalization of LV and aortic pressures at late diastole. All of these findings confirmed that late diastolic opening of the AV was the result of equalization of LV and aortic pressures because of markedly elevated LV EDP caused by AR. From the continuous-wave Doppler spectrum of AR, LV and aortic pressure waveforms were estimated as shown in Figure 7. LV EDP was estimated to be 44 mm Hg from the systemic diastolic pressure, and LV pre-A pressure was approximated as 17 mm Hg, which was calculated as the estimated LV EDP (44 mm Hg) minus the pressure gradient between the aorta and left ventricle just before atrial contraction (27 mm Hg). From these considerations, the hemodynamics of the patient were assumed to be slightly elevated mean left atrial pressure and severely elevated LV EDP, which could explain the absence of lung congestion despite markedly elevated LV EDP.

After the echocardiographic evaluation, computed tomography and carotid ultrasonography demonstrated wall thickening of the branches of the aorta (Figure 8). In addition to these findings, positive

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Conflicts of interest: The authors reported no actual or potential conflicts of interest relative to this document.

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https://doi.org/10.1016/j.case.2018.06.003

248-6441
HLA-B52 was revealed, and the patient was diagnosed with Takayasu’s arteritis, resulting in the initiation of steroid therapy thereafter.

**DISCUSSION**

Severe AR usually causes elevation of LV EDP and decline of systemic diastolic pressure, resulting in reduced pressure difference between the aorta and left ventricle at late diastole. However, equalization of the pressures of the two cavities rarely occurs. As far as we know, diastolic opening of the AV in AR has been reported in 10 articles (Table 1). It was first reported by Page and Layton in Marfan syndrome. Other conditions showing late diastolic opening of the AV were perforation of the leaflet secondary to infectious endocarditis, prolapse of the AV, and aortic dissection. However, this phenomenon has not been reported in Takayasu’s arteritis, and ours is the first report of late diastolic opening of the AV due to AR in Takayasu’s arteritis. Takayasu’s arteritis is a large-vessel vasculitis that mainly affects the aorta and its branch, resulting in stenosis, occlusion, or dilation of the vessels, and the dilation of the aortic root occasionally causes AR.

Prior reports concluded that the mechanism of diastolic opening of the AV is the equalization of aorta and LV pressures at late diastole because of marked elevation of LV EDP. They also reported that premature closure of the mitral valve due to the rapid rise in LV diastolic pressure was the second notable finding in these settings.
Consistent with these reports, we found signs of markedly elevated LV EDP suggested by the transmitral and pulmonary venous flow patterns along with late diastolic mitral regurgitation. In addition, we could further estimate the unique hemodynamics, relatively elevated mean left atrial pressure, and extremely elevated LV EDP from the continuous Doppler waveform of AR (Figure 6). These hemodynamics indicate that the regurgitant volume was large enough to mildly elevate the LV pre-A pressure and also to severely elevate LV EDP with the small increase in the volume during late diastole.

CONCLUSION

We report a case showing diastolic opening of the AV in functional severe AR due to a dilated aortic root associated with Takayasu’s arteritis, the hemodynamics of which could be precisely evaluated by comprehensive echocardiographic assessment.

ACKNOWLEDGMENT

We thank Yumi Takahashi and Hirokazu Komoriyama for their work managing the patient; Miwa Sarashina and Shingo Tsujinaga for their work collecting computed tomographic data; Masahiro Nakabachi for his work collecting echocardiographic data; Hisao Nishino, Shinobu...
Yokoyama, Kazunori Okada, Sanae Kaga, and Taisei Mikami for their work interpreting the data and for their advice about the discussion section; and Heffel Allen Paul for his work revising grammatical errors and sentence structure.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2018.06.003.

Figure 6  Continuous-wave Doppler image of AR showing extremely shortened pressure half-time (174 msec; dashed line) and abrupt decline of velocity just before atrial contraction to end-diastole (dashed circle) (A). Color M-mode Doppler image of AR showing discontinuation of regurgitation at late diastole (dashed circle) (B).

Figure 7  Estimated pressure waveforms of the left ventricle (LV) and aorta (A) and corresponding continuous-wave Doppler image of AR (B). Left ventricular EDP was estimated to be 44 mm Hg from the systemic diastolic blood pressure, and aortic–left ventricular pressure gradient just before atrial contraction was estimated to be 27 mm Hg from the continuous-wave Doppler image. Therefore, left ventricular pre-A pressure was estimated to be 17 mm Hg from their difference.
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Figure 8 Chest enhanced computed tomographic imaging showing wall thickening of the aorta from the ascending aorta, through the aortic arch, to the descending aorta and the pulmonary artery (A). Carotid ultrasonography showing diffuse and homogeneous wall thickening with medium echogenicity of the aortic arch branches (B). Mean and maximal intima-media thickness of left common carotid artery (L-CCA) were 0.24 and 0.27 cm, respectively (C). BCA, Brachial artery; R-CCA, right common carotid artery; R-SUBCLAV, right subclavian artery.

Table 1 List of previous reports of diastolic opening of AV

| Study | Etiology | Mechanism of AR | LV EDp (mm Hg) | DBP (mm Hg) |
|-------|----------|-----------------|----------------|-------------|
| Page and Layton (1975) | Marfan syndrome | Aortic root dilation | 50 | 50 |
| Weaver et al. (1977) | Infective endocarditis | Torn cusp | 54 | 54 |
| Pietro et al. (1978) | Rheumatic disease | Cusp perforation | 52 | 52 |
| Perez et al. (1978) | Unclear | Cusp fenestration | 50 | 50 |
| Cohen et al. (1979) | Rheumatic disease | Unclear | — | — |
| Nathan et al. (1982) | Infective endocarditis | Flail cusp | 54 | 46 |
| Martinez et al. (1986) | Marfan syndrome | Aortic root dilation | 63 | 60 |
| Meyer et al. (1987) | Sinus of Valsalva aneurysm, syphilitic aneurysm | Cusp prolapse | 43–82 | 41–82 |
| Shah et al. (2004) | Infective endocarditis | Large vegetation | — | 54 |
| Driocoll et al. (1981) | Marfan syndrome | Aortic dissection | 23 | — |
| Present case | Takayasu’s arteritis | Aortic root dilation | — | 44 |

DBP, Diastolic blood pressure.
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