Circulation Journal
Official Journal of the Japanese Circulation Society
http://www.j-circ.or.jp

Membranous-Type Supravalvular Aortic Stenosis in a 61-Year-Old Woman

Tsuyoshi Honda, MD; Hisanori Kanazawa, MD; Kazuteru Fujimoto, MD

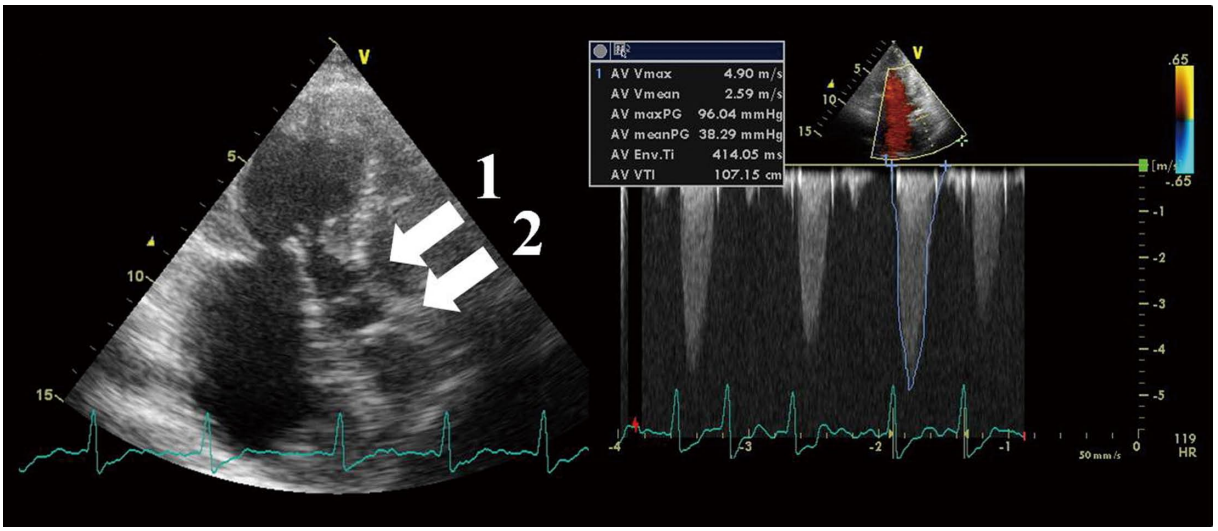

Figure 1. Transthoracic echocardiography indicated a tri-leaflet aortic valve with good opening of the leaflets (arrow 1) and a membrane-like structure (arrow 2) with a 94-mmHg peak pressure gradient above the aortic valve.

A 61-year-old woman was referred to Kumamoto Medical Center because of refractory heart failure. She had been well until the past year. Physical examination indicated a 4/6 ejection systolic murmur at the base of the heart radiating to both carotids. Electrocardiography suggested left ventricular hypertrophy. Transthoracic echocardiography showed a hypercontractile left ventricle, severe mitral regurgitation, a tri-leaflet aortic valve with good opening of the leaflets (arrow 1, **Figure 1**) and a membrane-like structure (arrow 2, **Figure 1**) with a 94-mmHg peak pressure gradient above the aortic valve (**Figure 1**). Diagnostic cardiac catheterization showed that there was no pressure gradient across the aortic valve, but a pressure gradient of 50–100 mmHg in the ascending aorta (**Figure 2**). Angiography showed a membranous structure at the sinotubular junction and a dilated ascending aorta (**Figure 2**). Coronary angiography showed no significant stenosis in any of the coronary arteries. Based on these examinations, the final diagnosis was a membranous-type supravalvular aortic stenosis (SVAS). The patient was managed by surgery of the aorta and the heart (supravalvular membrane resection and mitral valve replacement). Angiography of the pulmonary and renal arteries indicated no abnormalities. Although the patient had features suggestive of Williams-Beuren syndrome, her family did not consent to genetic analysis. After the 9-month follow-up visit, the patient was in good clinical condition.

Although the number of cases of adult congenital heart disease is increasing,[1] SVAS is an uncommon congenital cardiac anomaly characterized by varying degrees of left ventricular outflow tract obstruction beginning distal to the aortic valve. According to the previous report, three-quarters of the patients had segmental supravalvular narrowing, one-quarter had diffuse narrowing of the ascending aorta and few had the membranous type.[2] In addition, the prognosis of patients with SVAS is related to the severity of aortic stenosis, which, in turn, influences the age of presentation: accordingly, a 30-year survival is possible in only 12% of pediatric patients with severe SVAS.[3,4] The present case is interesting because of the occurrence of the

Received May 8, 2011; revised manuscript received June 14, 2011; accepted June 21, 2011; released online July 29, 2011 Time for primary review: 22 days
Department of Cardiology, Cardiovascular Center, National Hospital Organization Kumamoto Medical Center, Kumamoto, Japan
Mailing address: Tsuyoshi Honda, MD, PhD, Department of Cardiology, Cardiovascular Center, National Hospital Organization Kumamoto Medical Center, 1-5 Ninomaru, Kumamoto 860-0008, Japan. E-mail: thonda@kumamoto2.hosp.go.jp
ISSN-1346-9843 doi:10.1253/circj.CJ-11-0479

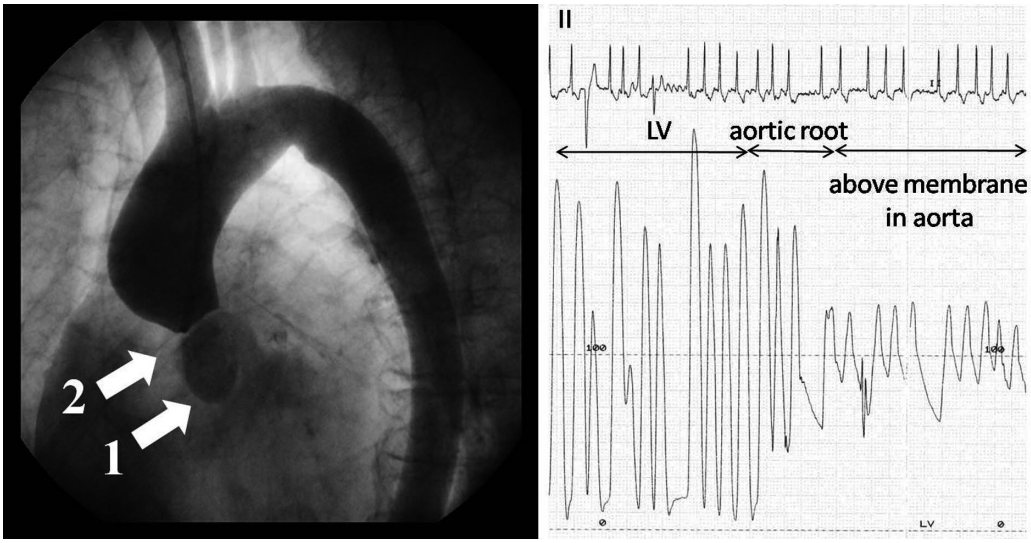

Figure 2. Aortic angiogram (left anterior oblique projection) showing the membranous stenosis of the ascending aorta (arrow 2) and aortic valve (arrow 1). Pullback pressure tracking from the left ventricle (LV) to the ascending aorta showed no pressure gradient across the aortic valve, but a pressure gradient of 50–100 mmHg across the supravalvular ascending aorta.

rare membranous type, the advanced age at presentation of symptoms and the long-term survival, despite the severity of aortic stenosis. Because this patient had hypertension and diabetes mellitus, these risk factors might exacerbate the severity of aortic stenosis as a result of atherosclerosis.

In the present case, echocardiography showed that the anterior mitral leaflet was thickening and shortening, and had prolapsed into the left atrium. Because the mitral regurgitation was not functional but valvular, mitral valve replacement was needed.[5]

Although uncommon, congenital membranes can present in adult life with clinical and echocardiographic features suggestive of aortic stenosis. Careful echocardiographic assessment should always be undertaken to confirm the precise diagnosis.

References

1. Toyoda T, Tateno S, Kawasoe Y, Shirai T, Shiina Y, Matsuo K, et al. Nationwide survey of care facilities for adults with congenital heart disease in Japan. *Circ J* 2009; **73:** 1147–1150.
2. Flaker G, Teske D, Kilman J, Hosier D, Wooley C. Supravalvular aortic stenosis: A 20-year clinical perspective and experience with patch aortoplasty. *Am J Cardiol* 1983; **51:** 256–260.
3. Kitchiner D, Jackson M, Walsh K, Peart I, Arnold R. Prognosis of supravalve aortic stenosis in 81 patients in Liverpool (1960–1993). *Heart* 1996; **75:** 396–402.
4. Keating MT. Genetic approach to cardiovascular disease: Supravalvular aortic stenosis, Williams syndrome, and long-QT syndrome. *Circulation* 1995; **92:** 142–147.
5. Komeda M, Kitamura H, Fukuya S, Okawa Y. Surgical treatment for functional mitral regurgitation. *Circ J* 2009; **73**(Suppl)**:** A-23–A-28.