A 42-year-old man was referred to our hospital for evaluation of systolic and diastolic murmurs at the left sternal border. Transthoracic 2D echocardiography revealed a quadricuspid pulmonary valve (Figure 1A) and a bicuspid aortic valve (Figure 1B). Transvalvular flow velocities at the pulmonary and aortic valves were 1.8 m/s and 1.7 m/s, respectively, suggesting mild pulmonary stenosis. Additionally, there was mild to moderate pulmonary and aortic regurgitation by color-Doppler. Magnetic resonance imaging also demonstrated these combined abnormalities of the semilunar valves (Figures 1C and 1D).

A quadricuspid pulmonary valve is rare, and the combination of a quadricuspid pulmonary valve and a bicuspid aortic valve is embryogenetically interesting. By the fourth week of gestation, a pair of bulbar ridges have formed in the cephalad portion of the truncus arteriosus. The semilunar valves are formed by mesenchymal outgrowth from the proliferations of the 2 bulbar ridges and the intercalated valvular swellings (Figure 2A). In this case, the abnormal cusp formations must have been embryologically caused by both the abnormal proliferations in the common trunk and aberrant fusion of the aortopulmonary septum (Figure 2B).

References

Figure 1. Two-dimensional echocardiograph of semilunar valves in systolic frame. Pulmonary valve (PV) with 4 cusps (A) and aortic valve (AV) with 2 cusps (B) are shown. Magnetic resonance imaging also demonstrated these combined abnormalities of pulmonary valve (C) and aortic valve (D).

Figure 2. Schematic diagrams of probable mechanisms of development of normal (A) and abnormal (B) semilunar valves. Combined abnormalities must have occurred both from abnormality of mesenchymal proliferation in common trunk and from abnormal septal division. Abbreviations as in Figure 1.
Combined Abnormalities of Semilunar Valves: Quadricuspid Pulmonary and Bicuspid Aortic Valves
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