A 35-year-old adult female was admitted to the hospital with dyspnea on exertion. The patient had no previously known heart disease and an unremarkable family history. Cardiac auscultation revealed a diastolic murmur at the second right intercostal space. Transthoracic echocardiography (TTE) revealed the almost complete absence of the posterior mitral leaflet (PML) and the bicuspid aortic valve (BAV). No clinical, microbiological, or imaging finding of infective endocarditis was observed. Subsequently, transesophageal echocardiography (TEE) was performed, which showed an elongated and mobile anterior mitral leaflet (AML) with severe hypoplasia of the PML (Figure A, Video A*) and the presence of a type 1 BAV with a raphe (Figure B, Video C*). A color Doppler ultrasound examination revealed mild mitral regurgitation and moderate aortic regurgitation. Three-dimensional TEE provided additional detail for the appraisal of the mitral valve abnormality (Figure C, D, Video B*). Ultimately, annual follow-up with TTE was suggested to the patient. Hypoplasia of the PML is an extremely rare congenital anomaly resulting from developmental arrest during conversion of the muscular chordae and leaflet to connective tissue. A dysplastic mitral valve coexisting with another cardiac abnormality is well known. However, a dysplastic mitral valve accompanied by BAV is unusual. In the literature, only 1 case report has suggested that both anomalies show familial aggregation, and thus, the condition may have a genetic basis. It is still not well known whether the association described in the present case is a genetic abnormality or just a coincidence. Accordingly, more investigation and experience are needed to answer this question about the relationship between hypoplasia of the PML and BAV.

**Figures**—(A) Transesophageal echocardiography (TEE) revealed a large, elongated anterior mitral leaflet with a mild degree of prolapse, severe hypoplasia of the posterior mitral leaflet (white arrow), and (B) the presence of a type 1 bicuspid aortic valve with a raphe. (C) Three-dimensional TEE short axis mid-systolic view of the mitral valve. (D) Mid-diastolic view.

*Supplementary video files associated with this presentation can be found in the online version of the journal.