A 59-year-old man with exertional dyspnea was referred for pre-operative evaluation before nephrectomy for a renal mass. Transthoracic echocardiography demonstrated Ebstein’s anomaly and marked left ventricular (LV) trabeculation (A and B, Online Videos 1, 2, and 3). Cardiac magnetic resonance imaging confirmed LV dilation with moderately to severely depressed global systolic function. There was extensive LV trabeculation, especially in the mid-ventricle and apex, and a mildly stenotic bicuspid aortic valve (BAV) (C to E, Online Videos 4 and 5). The maximal ratio of noncompacted to compacted myocardium was 2.8, consistent with the diagnosis of LV noncompaction. The displacement of the septal tricuspid leaflet from the mitral valve annulus was 1.9 cm (0.92 cm/m²).

The present case illustrates an unusual combination of left-sided abnormalities in a patient with Ebstein’s anomaly, including LV noncompaction, LV systolic dysfunction, and BAV. To our knowledge, this is the first reported case with this constellation of findings. LA = left atrial; RA = right atrial; RV = right ventricular.