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Coexistence of Left Ventricular Noncompaction and Aortic İnterruption

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A 22-year-old male patient was admitted to our hospital cardiology service suffering from dyspnea on exertion. On his physical examination, there were a 2/4 diastolic murmur in aortic valve area and a 3/6 systolic murmur both in the apical region and through the posterior chest wall. ECG showed normal sinus rhythm and left ventricular hypertrophy criteria. Real time two and three-dimensional transthoracic echocardiography (TTE) showed a mild regurgitant bicuspid aortic valve and a spongy appearance of the left ventricular wall (Fig. 1A–D).There were also dilatation of ascending aorta and the hypoplasia of the distal part of aortic arch and the descending thoracic aorta. In accordance with these TTE findings, contrast-enhanced magnetic resonance imaging (MRI) of the heart and the computerized tomography angiography (CTA) of the thoracic aorta was performed to demonstrate the pathology and to make a further evaluation. Spongy appearance of the left ventricular wall compatible with left ventricular noncompaction was seen in his contrast-enhanced cardiac MRI (Fig. 1E). We saw also regurgitation through the aortic valve during systolic phase in MRI CINE Sequence and diffuse myocardial hypertrophy of the left ventricle. Interruption at the level of distal aortic arch and hypoplasia of the descending thoracic aorta was seen in his CTA (Fig. 1F,G). Because the interruption is located distal to the left subclavian artery, it is a type A aortic interruption (AI) according to the classification by Celoria and Patton.[1] Dilatation of the ascending aorta and the collateral vascular structures were the additional findings in his CTA (Fig. 1H).

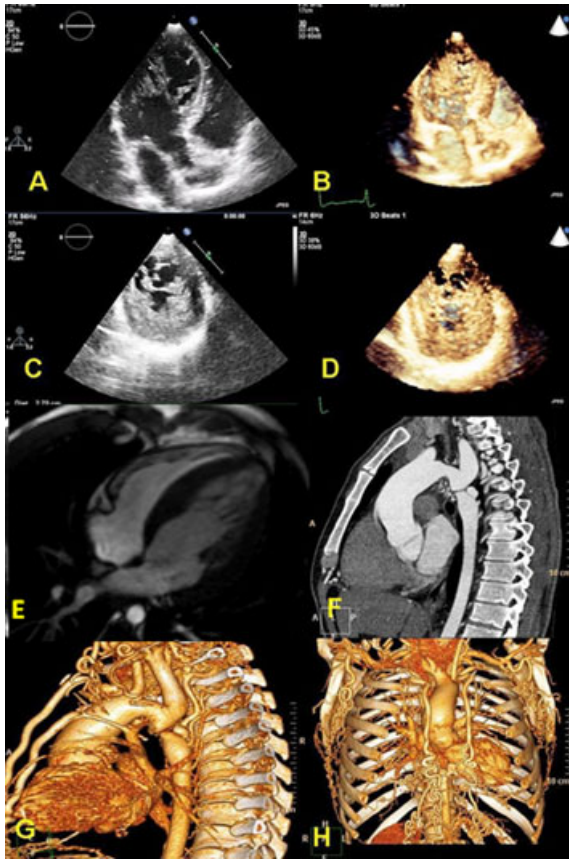

Figure 1. A., B., C., and **D.** Two- and three-dimensional transthoracic echocardiography (TTE) showed a spongy appearance of the left ventricular wall. **E.** Four-chamber MR image, spongy appearance of the left myocardial wall compatible with left ventricular noncompaction. **F.** Thoracic CT angiography, sagittal oblique MPR image. Interruption was seen at the distal part of the aortic arch. **G.** Thoracic CT angiography, sagittal oblique volume rendering image. Interruption was seen at the distal part of the aortic arch. **H.** Thoracic CT angiography, coronal volume rendering image. Multiple dilated arterial collateral vascular structures (especially look for both internal mammarian artery).

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AI is an extremely rare congenital anomaly. It occurs in 3 in every million live births, and accounts for 1% of all congenital heart disease.[2,3] Noncompaction of the ventricular myocardium is a cardiomyopathy thought to be caused by arrest of normal embryogenesis of the endocardium and myocardium and it is an uncommon finding.[4] AI is associated with an intra-cardiac malformation such as ventricular septal defect, patent ductus arteriosus, bicuspid aortic valve, left ventricular outflow tract obstruction or aortopulmonary window.[5] But our case is the first one in literature which the AI associated with left ventricular noncompaction.

References

1. Celoria GC, Patton RB: Congenital absence of the aortic arch. *Am Heart J* 1959;58:407–413.
2. Erden I, Kayapinar O, Erden EC, Yalçin S: Silent interrupted aortic arch in an elderly patient. *Cardiol J* 2011;18: 695–697.
3. Kleinrok A, Zaremba-Flis E, Smyk T: Interrupted aortic arch in an adult female. *Echocardiography* 2010;27: E70–E72.
4. Weiford BC, Subbarao VD, Mulhern KM: Noncompaction of the ventricular myocardium. *Circulation* 2004;109: 2965–2971.
5. Messner G, Reul GJ, Flamm SD, et al: Interrupted aortic arch in an adult: single-stage extra-anatomic repair. *Tex Heart Inst J* 2002;29:118–121.