Heterotaxy syndrome associated with left ventricular non-compaction

A 20-year-old asymptomatic man was referred to our clinic for the evaluation of a 2/6 grade systolic cardiac murmur. The ECG was normal. On his transthoracic echocardiography (TTE), subaortic membrane causing peak 25 mm Hg gradient on left ventricular outflow tract as the cause of the murmur (Fig. 1A) and an enlarged coronary sinus (CS) (Fig. 1B) were detected. We also suspected left ventricular non-compaction (Fig. 1C, D). We performed agitated saline injection via left antecubital vein for diagnosis of persistent left superior vena cava (PLSVC) and bubbles reached to CS before right atrium, indicating PLSVC. The injection was repeated via right antecubital vein to evaluate the right superior vena cava (RSVC) because of that the absence of RSVC may be associated with PLSVC. After the injection, the coronary sinus was opacified before right atrium, indicating the absence of RSVC. Left ventricular non-compaction was confirmed with cardiac magnetic resonance imaging (Fig. 2A, B). Thoracal computed tomographic (CT) venography was performed, and it confirmed the PLSVC and also revealed the absence of RSVC (Fig. 3A). Surprisingly, it also demonstrated absent inferior vena cava (IVC). Therefore, it forced us to perform abdominal CT angiography. It revealed the venous return of lower limbs and abdomen maintaining with dilated hemiazygous vein, indicating absent IVC. His abdominal CT faced us to another interesting findings, polysplenia (Fig. 3B).

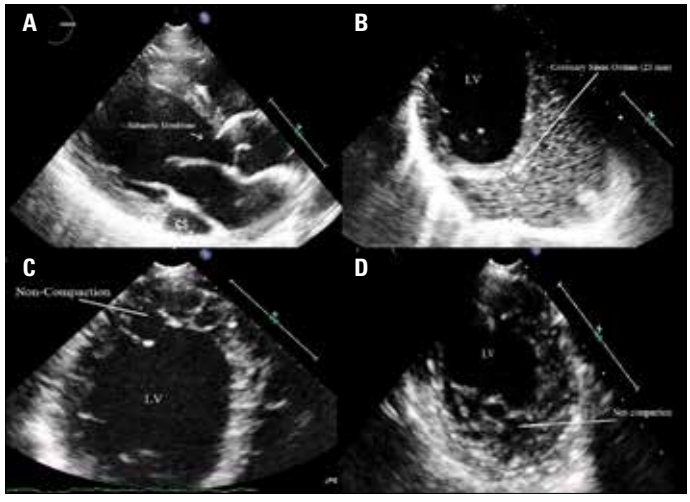

Figure 1. TTE. Subaortic membran and dilated CS (A), dilated CS ostium (B), LV non-compaction (C, D)

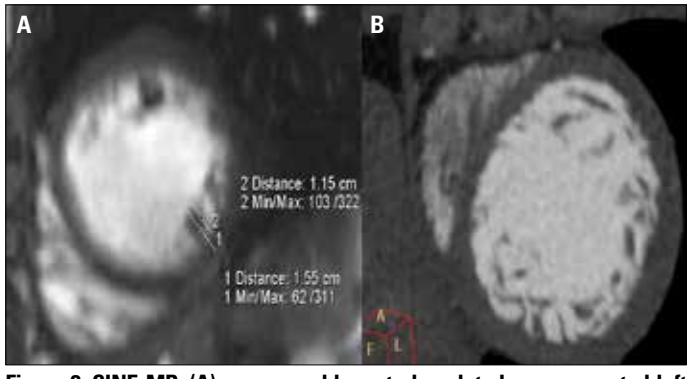

Figure 2. CINE-MR; (A) coarse and hypertrabeculated noncompacted left ventricle with a 2,8 non-compaction/compaction ratio. (B) MDCT; Prominent muscular trabeculations with deep intertrabecular recesses

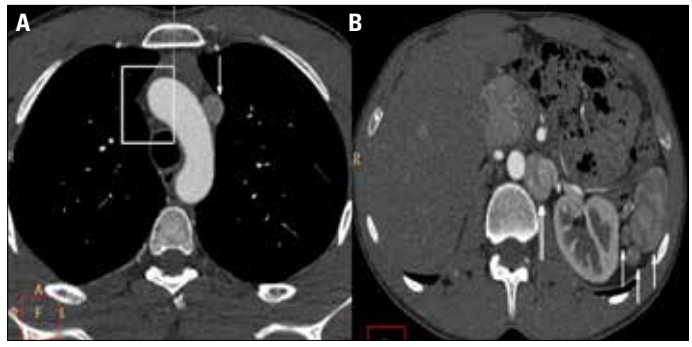

Figure 3. (A). Left-sided superior vena cava (arrow), the superior vena cava is absent on the right side (rectangle), (B). Multi dedector CT- Polysplenia (arrows) and hemiazygos continuation of the inferior vena cava (double arrow)

Subsequently, all findings in the entire story reached us to diagnose heterotaxy syndrome (HS). Coexistence of HS, subaortic membrane and left ventricular non-compaction has not been reported in the literature.

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Three-dimensional echocardiographic assessment of biatrial giant thrombi complicated with peripheral and pulmonary embolism

A 76-year-old woman admitted to the emergency department with complaint of right leg pain. She had hypertension, atrial fibrillation and she was bedridden since she had stroke 2 years ago. She was taking warfarin, amlodipine and valsartan therapy. On physical examination her lower extremity pulses were bilaterally not palpable. Doppler ultrasonographic examination revealed total occlusion of left main iliac and right superficial femoral arteries. Emergent catheter embolectomy for the right femoral artery was performed which resolved the ischemic symptoms. Enoxaparin 1mg/kg subcutaneously every 12 hours was initiated and oral warfarin 5 mg/day was started the following day. Transthoracic echocardiography and 2-D/3-D transesophageal echocardiography (TEE) showed a left atrial thrombus measuring 46x29 mm which was protruding to the mitral inlet (Fig.1, 2 and Video 1. See corresponding video/movie images at www.anakarder.com). Another thrombus measuring 25x23 mm was located in the right atrial appendage (Fig. 3, 4