

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).

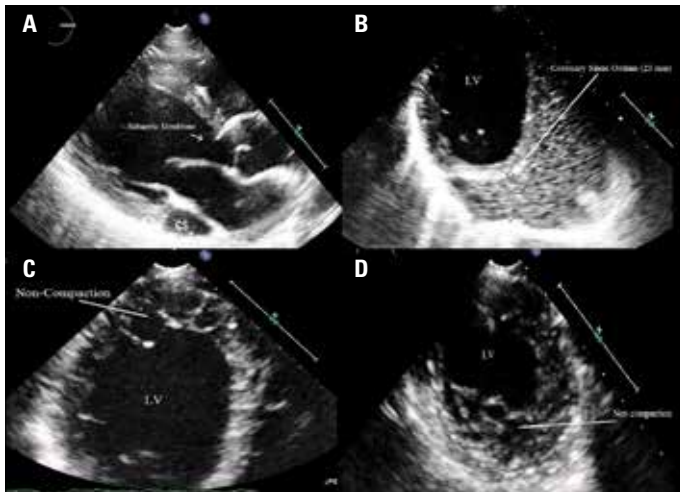


Figure 1. TTE. Subaortic membran and dilated CS (A), dilated CS ostium (B), LV non-compaction (C, D)

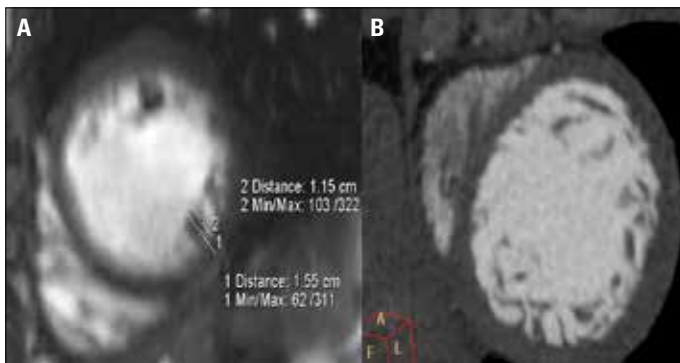


Figure 2. CINE-MR; (A) coarse and hypertrabeculated noncompacted left ventricle with a 2,8 non-compaction/compaction ratio. (B) MDCT; Prominent muscular trabeculations with deep intertrabecular recesses

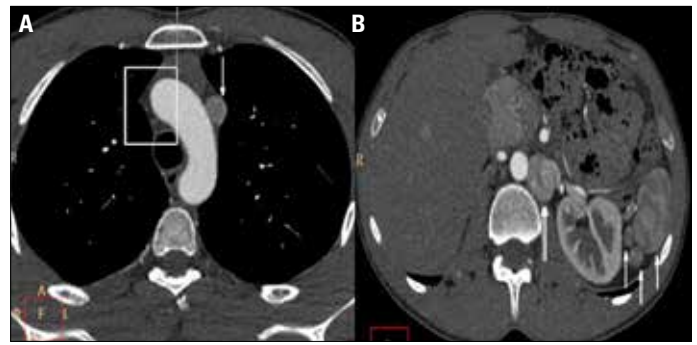


Figure 3. (A). Left-sided superior vena cava (arrow), the superior vena cava is absent on the right side (rectangle), (B). Multi dedector CT- Polysplenia (arrows) and hemiazygos continuation of the inferior vena cava (double arrow)

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

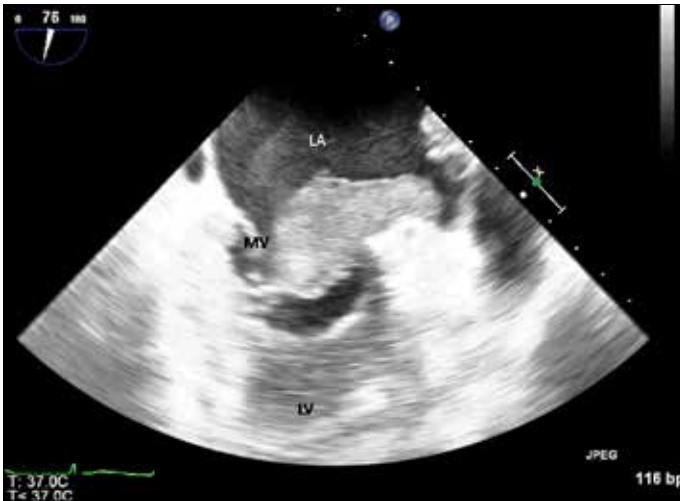


Figure 1. Two-dimensional transesophageal echocardiographic view showing large left atrial thrombus located at the left atrial appendage reaching the mitral inlet

LA - left atrium; LV - left ventricle; MV - mitral valve

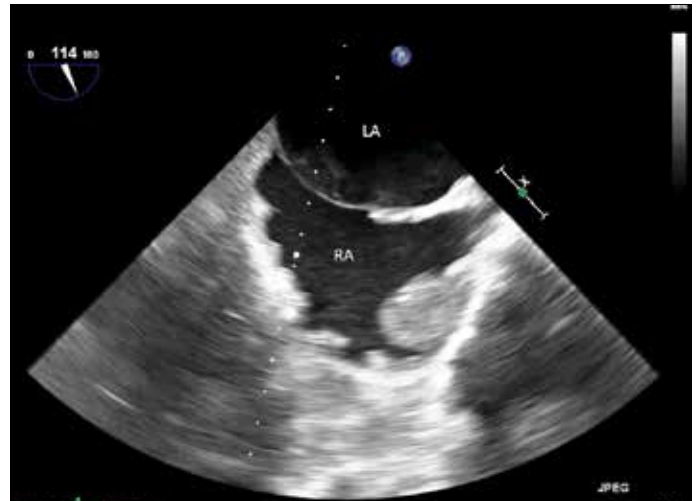


Figure 3. Two-dimensional transesophageal echocardiographic view showing right atrial thrombus located at the right atrial appendage

LA - left atrium; RA - right atrium

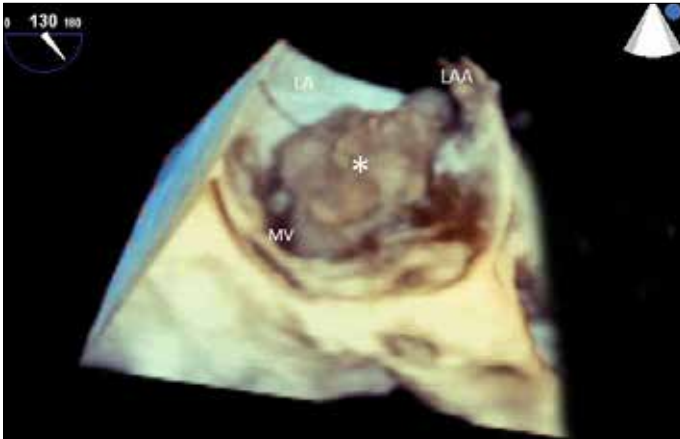


Figure 2. Three-dimensional transesophageal echocardiographic view showing large left atrial thrombus (asterisk) located at the left atrial appendage reaching the mitral inlet

LA - left atrium; LAA - left atrial appendage; MV - mitral valve



Figure 4. Three-dimensional transesophageal echocardiographic view showing right atrial thrombus (asterisk) located at the right atrial appendage

CT - crista terminalis; RA - right atrium; SVC - superior vena cava

and Video 2. See corresponding video/movi images at www.anakarder.com). Computed tomography angiography revealed biatrial thrombi, bilateral filling defects in the pulmonary arteries compatible with pulmonary embolism and total occlusion of left main iliac artery (Fig. 5). Right femoral artery was patent. Surgical thrombectomy of the atrial thrombi was suggested however the patient refused the operation. Thus, the patient was discharged after optimal anticoagulation with warfarin treatment was achieved. The patient was called for re-evaluation but she was lost to follow-up.

Transesophageal echocardiography is essential in diagnosis of embolism of cardiac origin. Biatrial giant thrombi complicated by multi-site embolism is a rare clinical finding. 3-D echocardiography imaging may yield superior images of the thrombi compared to 2-D imaging.

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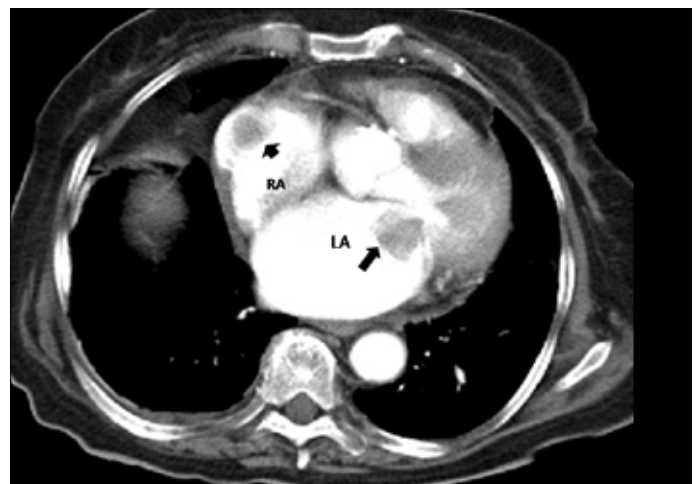


Figure 5. Computed tomography of the chest with contrast showing left (arrow) and right atrial (short arrow) thrombi

LA - left atrium; RA - right atrium

Video 1. Three-dimensional transesophageal echocardiographic view of the large mobile left atrial thrombus

Video 2. Three-dimensional transesophageal echocardiographic view of the right atrial thrombus located at the right atrial appendage

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Real-time monitoring of the giant right atrial thrombus prolapsing into the right ventricle and the deterioration of the thrombus with thrombolytic treatment by transthoracic echocardiography

A 57-year-old male with a history of metastatic pancreatic adenocarcinoma presented to our clinic with complaints of dyspnea for seven days. Arterial blood pressure and heart rate were 100/60 mm Hg and 116 bpm respectively. Electrocardiography showed S1-Q3-T3 pattern with mild sinus tachycardia. Two-dimensional transthoracic echocardiography (TTE) revealed a giant, highly mobile thrombus in the right atrium prolapsing into right ventricle through the tricuspid orifice during diastole (Fig. 1, Video 1. See corresponding video/movie images at www.anakarder.com). Enlarged right heart chambers, moderate tricuspid regurgitation and elevated (95 mm Hg) pulmonary artery systolic pressure were also noted. Ejection fraction was 60%. Contrast-enhanced 64-slice computed tomography demonstrated bilateral central pulmonary embolism (PE) with giant right atrial thrombus (Fig. 2). We decided to administer intravenous thrombolytic therapy under the diagnosis of right heart thrombosis with massive PE and he was given 100 mg of tissue-type plasminogen activator (t-PA) over two hours. After initiating thrombolytic infusion, the patient underwent TTE for 10 minutes each. The deterioration of the giant thrombus was observed in real-time (Fig. 3, Video 2. See corresponding video/movie images at www.anakarder.com). His symptoms completely resolved and he was discharged from the hospital after five days.

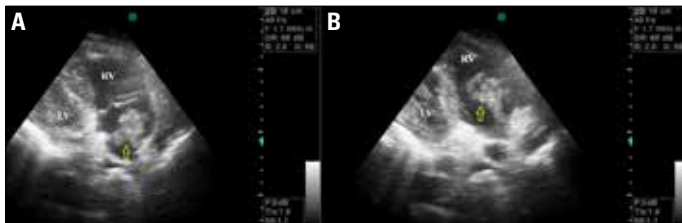


Figure 1. (A) Apical 4-chamber view of the giant right atrial thrombus during systole (yellow arrow) (B) Apical 4-chamber view of the giant right atrial thrombus (prolapsing into right ventricle through tricuspid orifice) during diastole (yellow arrow)

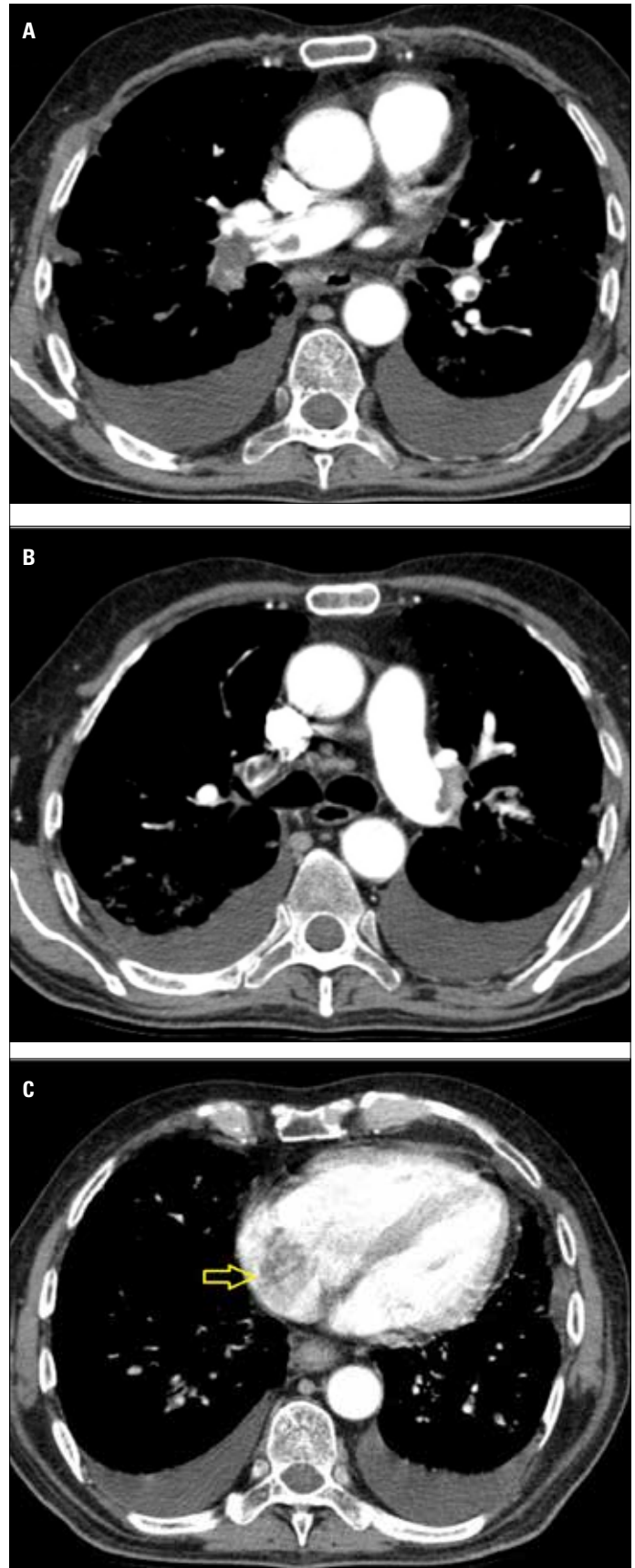


Figure 2. (A-C) Contrast-enhanced 64-slice computed tomography showing bilateral pulmonary embolism and giant right atrial thrombus (yellow arrow)