CASE IMAGE

Unusual case of tricuspid atresia and transposition of the great arteries presenting with aortic dissection: A rare condition

Olağandışı triküspit atrezi olgusu ve aort diseksiyonu ile ortaya çıkan büyük arterlerin transpozisyonu: Nadir bir durum

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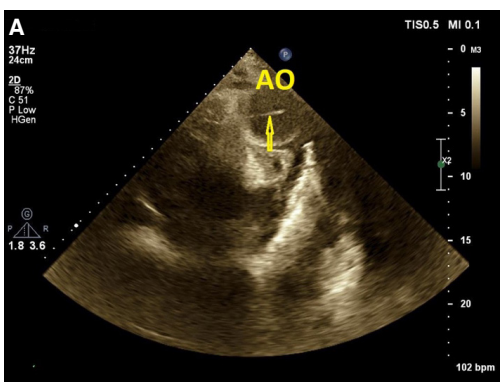
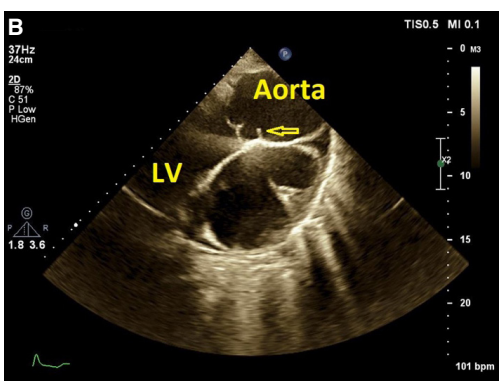
A 19-year-old female was referred to the emergency department with severe dyspnea, chest pain, and orthopnea. At the time of arrival, her vital signs were a blood pressure of 160/90 mm Hg, a pulse rate of 109 beats/minute, a respiratory rate of 30 breaths/minute, and an oxygen saturation rate of 82% at room temperature. In the preliminary evaluation, the patient's lips and fingertips were cyanotic and digital clubbing was detected. Chest auscultation revealed bilateral rales in the lower third. Pitting edema measuring 1+ was observed in the lower extremities. Detailed echocardiography revealed abdominal situs inversus, inferior vena cava continuity to the right atrium, a rightward apex suggestive of dextrocardia, L-type ventricular looping, an ambiguous atrioventricular connection (univentricular atrioventricular connections), and a discordant ventriculoarterial connection. The pulmonary artery was on the left posterior side of the aorta, originating from the morphological left ventricle (LV) and the aorta originated from the morphological right ventricle. The LV was severely enlarged and demonstrated impaired systolic function (LV ejection fraction: 30%). The tricuspid valve was atretic. In addition, a right-sided aortic arch; severe mitral regurgitation; severe aortic insufficiency; severe valvular and subvalvular pulmonary stenosis; a large, outlet-type ventricular septal defect (22 mm), a large atrial septal defect (35 mm) with a bidirectional shunt, and an aneurysmal ascending aorta (56 mm) were observed. The pulmonary artery (PA) and its branches were well-developed (main PA: 23 mm, right PA: 16 mm, left PA: 11 mm), but there was a dissecting flap originating from the posterior wall of the aorta above the sinotubular junction that indicated a type A aortic dissection (Fig. A, B and Video 1*). Based on the clinical findings, the patient was diagnosed with aortic dissection type A and pulmonary edema. Treatment with intravenous furosemide and nitroglycerin was initiated. Following medical treatment, the signs of pulmonary edema of orthopnea, dyspnea, and rales ameliorated, and the patient's general condition improved. The condition of the patient was discussed by a team in a heart conference, but the patient did not consent to further treatment and left the hospital. After 6 months of follow up the patient survived, but continued to experience dyspnea. There is no direct evidence of an association between aortic dilatation and dissection with an unrepaired transposition of the great arteries (TGA). The co-existence of anomalies such as tricuspid atresia, pulmonary stenosis, ventricular septal defect, atrial septal defect, and TGA is extremely rare, and aortic dissection has not previously been reported in this patient population, to our knowledge. This case emphasizes the necessity of close, life-long follow-up in adolescent patients with a congenital heart defect due to the greater risk of arterial complications, such as a dissection.





Figures– (A, B) Transthoracic echocardiography images demonstrate the dissecting flap (arrow) in the dilated ascending aorta, originating from the posterior wall of the aorta above the sinotubular junction. AO: Aorta; LV: Left ventricle.

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