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Hypoplastic aorta in a patient with familial hypercholesterolemia

Ailesel hiperkolesterolemili bir hastada hipoplastik aorta

A 20-year-old man was admitted with extensive lesions on his hands. He had cutaneous xanthomas on the back of the hands, knees and elbows (Fig. 1A). Total cholesterol, low and high density lipoprotein cholesterol (LDL-C and HDL-C) were 626 mg/dL, 536 mg/dL and 74 mg/dL respectively and familial hypercholesterolemia (FH) was diagnosed. Transthoracic echocardiography (TTE) revealed degenerative changes in the aortic valve and mild aortic regurgitation. Computed tomography (CT) showed diffuse calcium plaques in the thoracic aorta (Fig. 1B, C). Abdominal aorta was 9.6 mm from hiatus to renal artery origin, 13.2 mm distal to this segment and 15.1 mm proximal to this segment (Fig. 1D). FH is an autosomal dominant disorder. Clinically, this is manifested as tendinous xanthomata and premature atherosclerosis. Hypoplasia of the aorta is a rare entity comprising tubular hypotrophy of a large segment of the thoracic and the abdominal aorta. Patients with hypoplasia of the infrarenal aorta is increased the incidence of atherosclerosis. Although hypoplasia of the abdominal aorta accompanied by FH may seem coincidence, we showed this condition because of its relationship early atherosclerosis. TTE is the first-step modality for cardiovascular imaging in adults with heart disease. The windows of access with transthoracic echocardiography may be inadequate for all regions of interest. Therefore, the patients with FH should be evaluated the further imaging such as CT and magnetic resonance imaging for the development of early atherosclerosis.

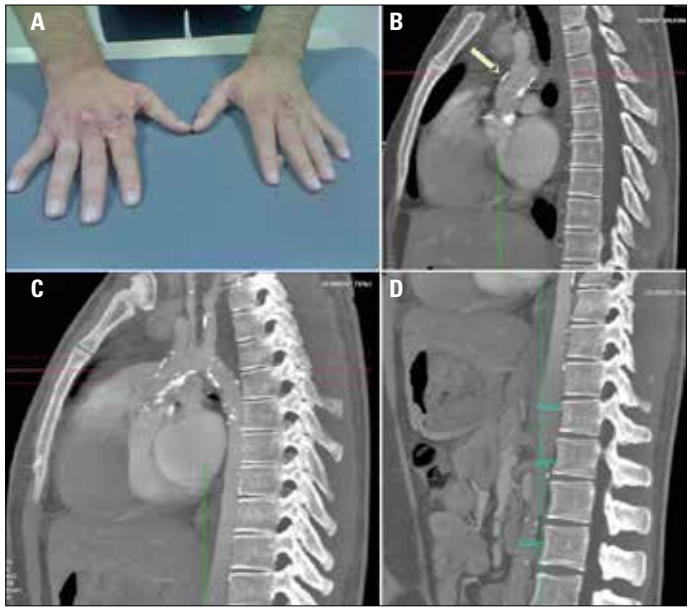

Figure 1. Xanthomas on the back of the his hands (A), computed tomography oblique sagittal images showed diffuse calcium plaques in the thoracic aorta especially in the ascending aorta (B, C), abdominal aorta was 9.6 mm from hiatus to renal artery origin, 13.2 mm distal to this segment and 15.1 mm proximal to this segment (D)

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A case of malposition of ventricular electrode through atrial septal defect

Atriyal septal defekt yoluyla uygunsuz yerleştirilen ventriküler elektrot olgusu

A 39-year-old man, who had a single-chamber pacemaker implanted for symptomatic bradycardia six years ago, was admitted because of dizziness and pre-syncope. Electrocardiogram showed sinus rhythm with right bundle brunch block and the chest X-ray demonstrated a pacemaker and its single electrode (Fig. 1). When pacemaker control was performed, the ventricular threshold was much higher than expected. Transthoracic echocardiography (TTE) revealed dilated right heart chambers, moderate tricuspid regurgitation and elevated pulmonary arterial pressure. In addition, an abnormal route of the ventricular electrode from the right atrium to the left atrium through atrial septal defect (ASD) was seen (Fig. 2A, Video 1. See corresponding video/movie images at www.anakarder.com). Transesophageal echocardiography (TEE) also demonstrated that

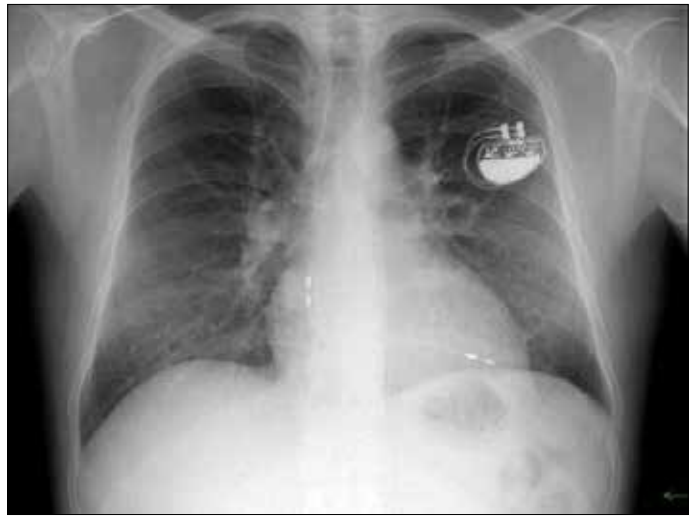

Figure 1. Chest X ray shows pacemaker and electrode

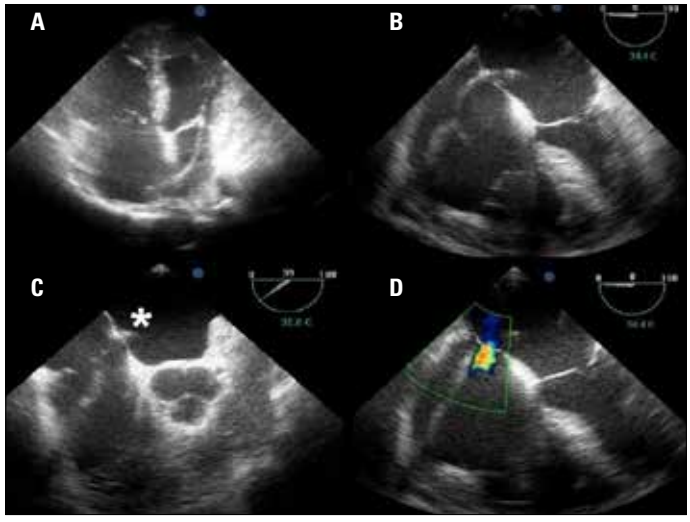

Figure 2. (A) The route of ventricular electrode from the right atrium to the left ventricle on TTE; (B) Ventricular lead passes ASD and reaches to the left atrium on TEE, four chamber view; (C) Ventricular electrode passes ASD (asterisk) on TEE, short-axis view of the aortic valve; (D) Transition from ASD by color Doppler and the position of the electrode
ASD - atrial septal defect, TEE - transesophageal echocardiography

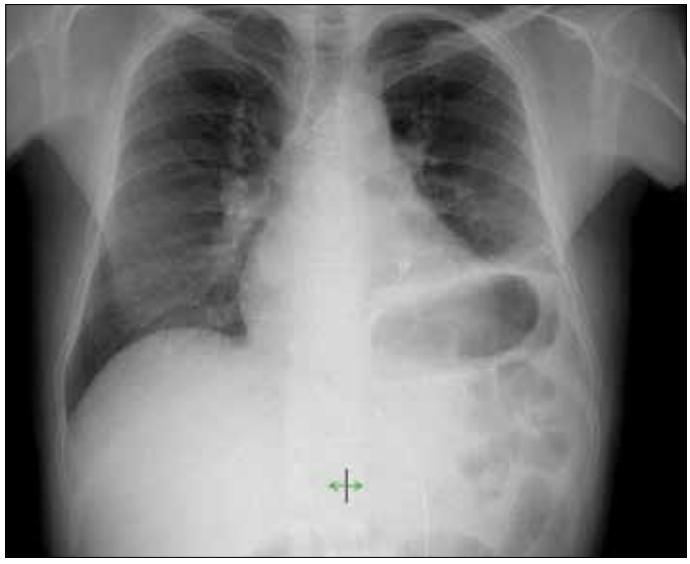

Figure 3. Chest X ray after surgery shows epicardial electrode

ventricular electrode reached to the left atrium via ASD (Fig. 2B-D, Video 2. See corresponding video/movie images at www.anakarder.com). Subsequently, the electrode passed the mitral valve posterior leaflet and reached the left ventricular lateral wall (Video 3. See corresponding video/movie images at www.anakarder.com). Neither vegetation nor thrombus was detected around the electrode. The patient was referred to the cardiac surgery and surgical removal of the malpositioned electrode, closure of the ASD, repair of the damaged mitral valve and implantation of abdominal pacemaker with epicardial electrode were performed (Fig. 3). His further clinical course was uneventful.

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Video 1. The route of malpositioned electrode on TTE
TTE - transthoracic echocardiography
Video 2, 3. Ventricular electrode passes ASD and mitral valve on TEE
ASD - atrial septal defect, TEE - transesophageal echocardiography

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Bivalvular calcification in a 9-year-old child presenting with syncope

Senkop ile başvuran 9 yaşındaki hastada bivalvüler kalsifikasyon

A 9-year-old girl was admitted to our outpatient with complaints of syncope following exertion. The patient had a history of six glaucoma surgeries. Echocardiography identified a thick anterior mitral valve leaflet with hyperechogenicity. A diastolic gradient with a maximum of 10 mm Hg and an average of 4.6 mm Hg was measured between the left atrium-left ventricle, which demonstrated restricted movement (Video 1. See corresponding video/movie images at www.anakarder.com). In the parasternal short-axis cross-section, aortic valve cusps were observed as being thick and hyperechogenic with restricted movement (Video 2. See corresponding video/movie images at www.anakarder.com). A thick, calcific, hyperechogenic abnormal chord structure was observed on the outflow tract of the left ventricle, extending to the outflow tract of the mitral posterolateral leaflet chord. Color Doppler examination revealed turbulent aortic flow. With CW Doppler, a systolic gradient of a maximum of 123 mm Hg, with average of 67 mm Hg, was identified between the left ventricle and aorta. It was observed that the mitral valve anterior leaflet, the aortic annulus, and the endo-

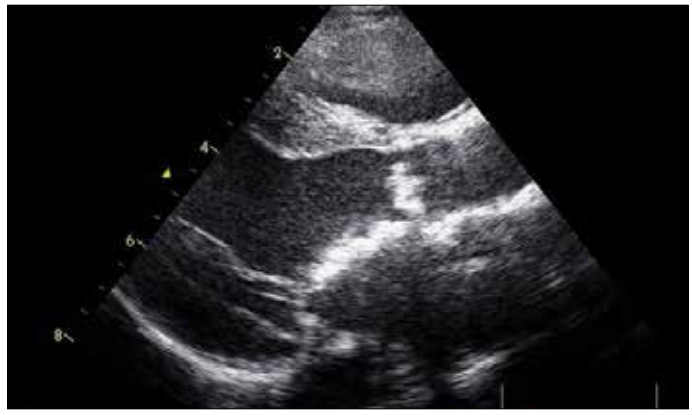

Figure 1. In the parasternal long axis cross-section, anterior mitral valve leaflet, the aortic annulus, and the endocardium layer were thick and hyperechogenic