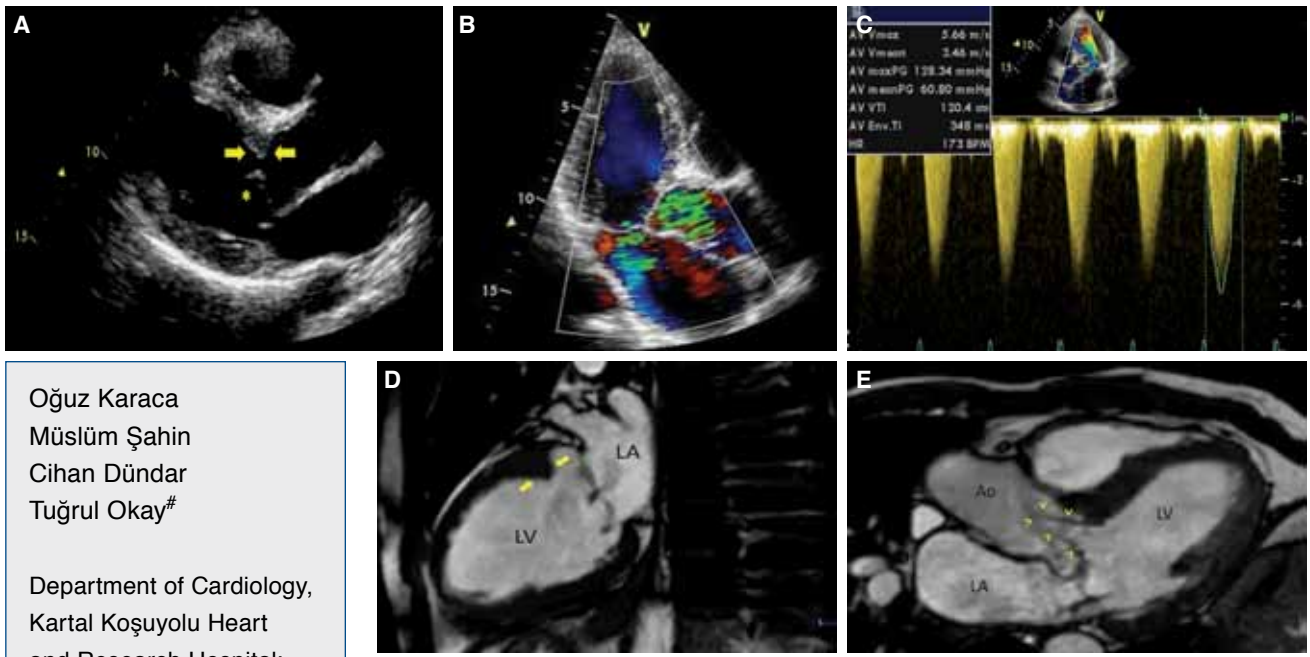
Unusual hypertrophic cardiomyopathy and extremely elongated chordae tendineae causing significant left ventricular outflow tract obstruction

Ciddi sol ventrikül çıkım yolu tıkanıklığına yol açan aşırı uzun korda tendinea ve hipertrofik kardiyomiyopati



Oğuz Karaca
Müslüm Şahin
Cihan Dündar
Tuğrul Okay#

Department of Cardiology, Kartal Koşuyolu Heart and Research Hospital; #Department of Cardiology, Yeşilköy International Hospital, both in İstanbul

A 36-year-old man presented with exertional dyspnea and recurrent episodes of syncope. He had no past history of coronary artery disease nor any risk factors for atherosclerosis. His blood pressure and heart rate were normal. Auscultation revealed a systolic thrill over the lower left sternal border and a grade 4/6 systolic ejection murmur. Electrocardiography showed prominent Q waves in V4-6 and nonspecific ST-T abnormalities in anterior leads. Transthoracic echocardiography revealed hypertrophy of the basal interventricular septum protruding into the left ventricular outflow tract (LVOT) and bileaflet mitral valve prolapse with systolic anterior motion of an elongated chorda (Fig. A). Septal wall thickness was 21 mm at the most prominent site. Other myocardial regions had normal wall thickness. Color Doppler analysis showed a systolic flow turbulence with a pressure gradient of 128 mmHg through the LVOT and posteriorly directed mitral regurgitation of moderate degree (Fig. B, C). Transesophageal echocardiography confirmed prominent septal hypertrophy narrowing the distal portion of the LVOT and systolic anterior motion of the hyperelongated chorda causing prolapse of the anterior and posterior mitral valve leaflets into the left atrium. Cardiac magnetic resonance imaging better visualized the pyramid-shaped septal hypertrophy (Fig. D), floating-like chordal elongation and its prolapse through the aortic valve (Fig. E), and bileaflet mitral valve prolapse. Localized late gadolinium enhancement was also noted confirming myocardial fibrosis at the area of septal hypertrophy. Left heart catheterization showed normal coronary arteries with a significant pressure gradient of 112 mmHg between the aorta and the LVOT. The patient underwent septal myectomy and mitral valve repair with chordal shortening. Following surgery, the pressure gradient decreased to 25 mmHg with minimal mitral regurgitation and marked improvement in the symptoms.

Figures. Transthoracic echocardiograms showing **(A)** localized basal septal hypertrophy protruding into the LVOT (arrows) and systolic anterior motion of the elongated chorda (asterisk) (parasternal long-axis view) and **(B)** systolic turbulent flow through the LVOT and posteriorly directed mitral regurgitation jet (apical long-axis view). **(C)** Continuous-wave Doppler showing a pressure gradient of 128 mmHg through the LVOT. Cardiac magnetic resonance images demonstrating **(D)** asymmetric upper septal hypertrophy protruding into the LVOT (arrows) with normal wall thickness of the remaining left ventricle and **(E)** extremely elongated chordae tendineae (arrow heads) prolapsing through the aorta.