Supravalvular aortic stenosis secondary to severe lipid accumulation in the ascending aorta in a patient with uncontrolled familial hyperlipidemia

Kontrol edilmemiş ailesel hiperlipidemi olan bir hastada, çıkan aortta ciddi lipit birikmesine bağlı olarak kapak seviyesi üzerinde gelişen aort darlığı

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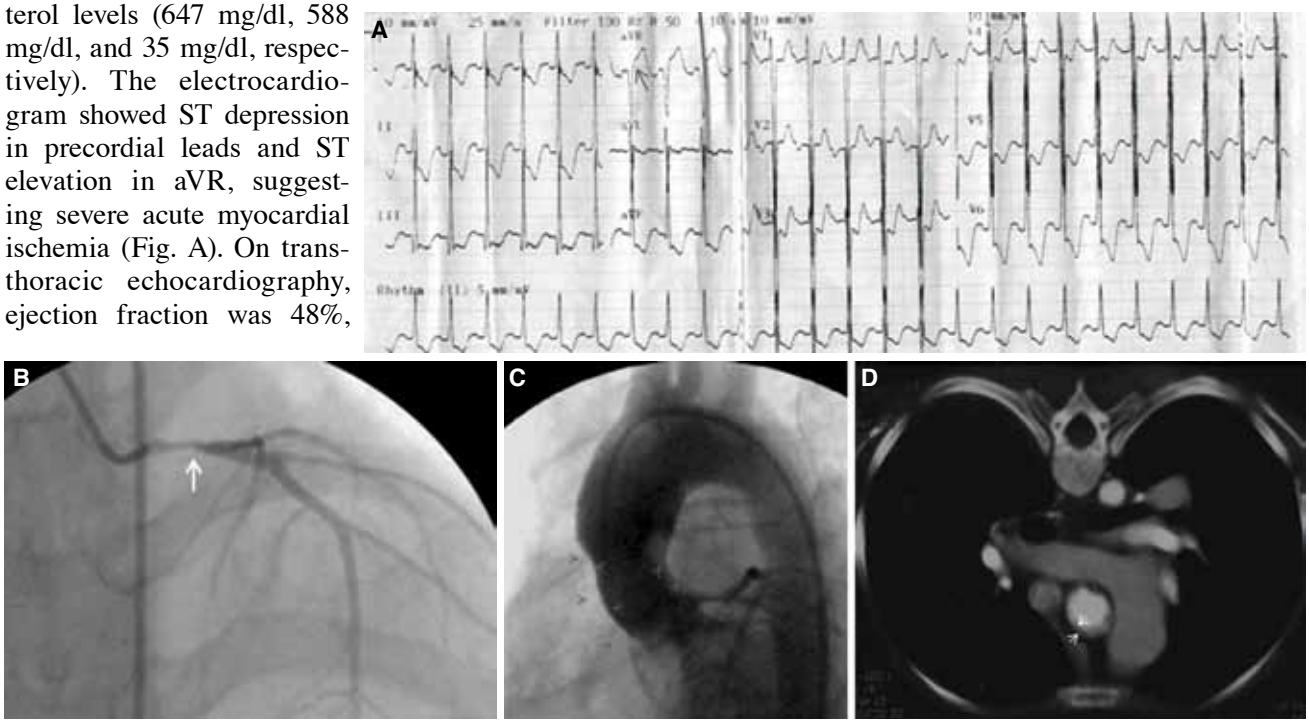
A 13-year-old girl with a previous diagnosis of type II hyperlipidemia was admitted with severe retrosternal chest pain. She had typical stigmas of familial hyperlipidemia including tendinous, tuberous and plantar xanthoma, and arcus cornealis. Cardiac examination showed sinus tachycardia of 115 bpm, blood pressure of 85/55 mmHg, and systolic and early diastolic murmurs. Serum cardiac markers were elevated (CK-MB 83 U/l, troponin I 5.73 ng/dl). Lipid profile showed increased total cholesterol and LDL cholesterol and decreased HDL cholesterol levels (647 mg/dl, 588 mg/dl, and 35 mg/dl, respectively). The electrocardiogram showed ST depression in precordial leads and ST elevation in aVR, suggesting severe acute myocardial ischemia (Fig. A). On transthoracic echocardiography, ejection fraction was 48%, and there were left ventricular dilatation, moderate mitral regurgitation, and a systolic aortic pressure gradient. Transesophageal echocardiography showed hypokinesia of the anteroseptal wall, supraannular aortic narrowing, and mild aortic valvular restriction. Color Doppler examination showed mild aortic and moderate mitral regurgitation. Coronary angiography revealed total occlusion of the ostial segment of the right coronary artery and a critical lesion (%50-60) in the distal left main coronary artery (Fig. B). Conventional aortography (Fig. C, Video file*) and computed tomography (Fig. D) showed severe aortic mural lipid accumulation. At emergency operation, the ascending aorta was dilated with a composite graft and coronary artery bypass operation was performed with grafting of the left anterior descending and circumflex arteries. Despite hemodynamic stability, the patient developed respiratory insufficiency due to aspiration pneumonia on the second postoperative day and died the following day.



Figures. (A) Electrocardiogram showing ST depression in the precordial leads and ST elevation in the aVR (arrow). **(B)** Coronary angiography showing a critical lesion (%50-60) in the distal left main coronary artery. **(C)** Aortography and **(D)** contrast-enhanced computed tomography images showing severe lipid accumulation (arrows). **Supplementary video file (Aortography in the right anterior oblique position) can be found in the online version.*