

Two-in-One: single coronary ostium and mitral valve prolapsus in a young female with Alport syndrome



İkisi bir arada: Alport Sendromu olan genç kadın hastada tek koroner ostiyum ve mitral kapak prolapsusu

A 23-year-old female with chronic renal failure because of Alport syndrome (AS) consulted for cardiac evaluation before renal transplantation. She described dyspnea with minimal effort and atypical chest pain. Past medical history includes adulthood onset, autosomal recessive type AS, due to a missense mutation in the COL4A3 gene, with development of severe renal insufficiency, hypertension, anterior lenticonus and mild sensorineural deafness for 4 years. She was taking carvedilol and amlodipine for hypertension. Examination showed arrhythmic pulse, apical 3/6 systolic murmur, other systems and biochemical parameters were unremarkable except renal function tests. Electrocardiogram revealed atrial fibrillation. Transthoracic echocardiography (TTE) revealed left ventricular (LV) ejection fraction of 55%, LV end-diastolic diameter of 59 mm, prolapsus of the posterior mitral leaflet and severe mitral regurgitation (MR) (Fig. 1A, 1B, Video 1, 2. See corresponding video/movie images at www.anakarder.com). Further investigation with transesophageal echocardiography (TEE) disclosed prolapse of the posterior mitral leaflet with severe eccentric mitral insufficiency jet flow directing to opposite site of effected leaflet and also no characteristic features for single mitral

orifice. Coronary angiography demonstrated both left main coronary artery (LMCA) and right coronary artery (RCA) were originating from the right sinus of Valsalva (RSV) via single ostium (Fig. 1C). Coronary system was free of atherosclerosis except 30% stenosis at proximal RCA. The LMCA was oriented retro-aortic and coursed down as LAD in the interventricular groove after giving rise to intermediate and circumflex arteries (Lipton RI-P) (Fig. 1D). Left ventriculography confirmed severe mitral regurgitation. Therefore, the patient was referred for mitral repair before renal transplantation.

AS is a rare inherited disorder characterized by involvement of the kidneys because of the defect in the genes encoding a connective tissue protein, one of several subunits of collagen (particularly type IV) ultimately leading to renal failure at an early age (1). Cardiac involvement has been reported rarely which most commonly includes conduction system abnormalities. Valvular and coronary anomalies in AS have not been reported previously. In our case, concomitant occurrence of previously unreported AS with MVP may be due to mutation at the level of collagen synthesis (2). In addition, single coronary ostium may contribute to this association coincidentally.

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Video 1: Apical 4-chamber echocardiographic views of posterior leaflet prolapsus

Video 2: Color Doppler apical 4-chamber view showing severe mitral regurgitation (eccentric)

References

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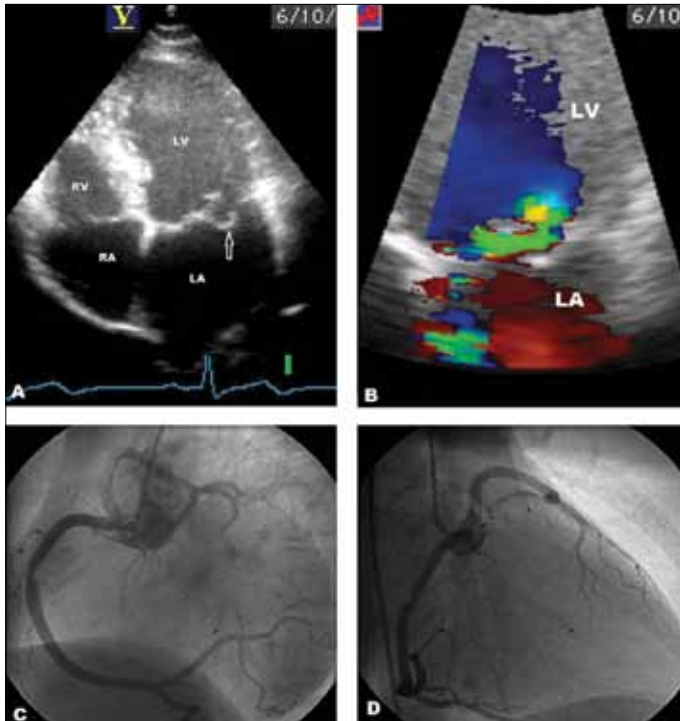


Figure 1. A) TTE showing prolapsus of the posterior mitral leaflet, **B)** Color Doppler showing eccentric severe MR, **C)** Coronary angiography revealed single coronary ostium (left anterior oblique view), **D)** LMCA coursed down as LAD in the interventricular groove after giving rise to intermediate and circumflex arteries (Lipton RI-P) (right anterior oblique view)

LAD - left anterior descending artery, LMCA - left main coronary artery, MR - mitral regurgitation, TTE - transthoracic echocardiography

Amiodarone-induced pleural fluid is not always accompanied by a risk factor

Amiodarona bağlı plevral sıvı: Her zaman eşlik eden bir risk faktörü olmayabilir

A 39-year-old male patient was hospitalized in Cardiology Department with Brugada syndrome in 2007 and DDDR/ICD (dual chamber rate-adaptive pacemaker/ implantable cardioverter-defibrillator) was implanted. Amiodarone in a dose of 200 mg/day was started. Mild pericardial effusion was detected in February 2010 and regressed in 2 weeks with indomethacin. He was hospitalized with pleural effusion in Department of