

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.

Uğur Canpolat, Kudret Aytemir, Lale Tokgözoğlu
Department of Cardiology, Faculty of Medicine, Hacettepe University, Ankara-Turkey

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

1. Alport AC. Hereditary familial congenital haemorrhagic nephritis. Br Med J 1927; 1: 504-6. [CrossRef]
2. Bassareo PP, Marras AR, Mercurio G. Ventricular septal defect in a child with Alport syndrome: a case report. BMC Cardiovasc Disord 2010; 10: 48. [CrossRef]

Address for Correspondence/Yazışma Adresi: Dr. Uğur Canpolat
Hacettepe Üniversitesi Tıp Fakültesi, Kardiyoloji Anabilim Dalı, 06100 Sıhhiye,
Ankara-Türkiye

Phone: +90 312 305 17 80 Fax: +90 312 305 41 37

E-mail: dru_canpolat@yahoo.com

Available Online Date/Çevrimiçi Yayın Tarihi: 13.03.2012

©Telif Hakkı 2012 AVES Yayıncılık Ltd. Şti. - Makale metnine www.anakarder.com web sayfasından ulaşılabilir.

©Copyright 2012 by AVES Yayıncılık Ltd. - Available on-line at www.anakarder.com
doi:10.5152/akd.2012.080

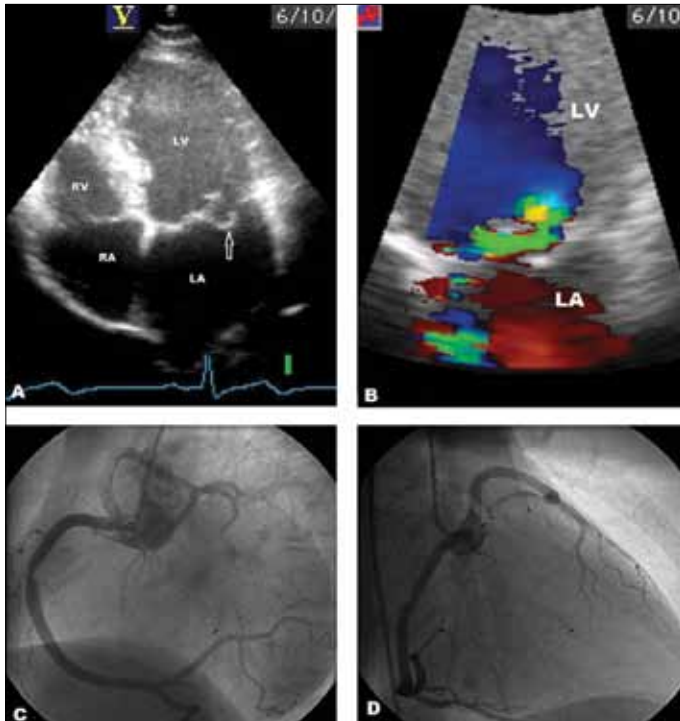


Figure 1. A) TTE showing prolapsus of the posterior mitral leaflet, **B)** Color Doppler showing eccentric severe MR, **C)** Coronary angiography revealed single coronary ostium (left anterior oblique view), **D)** LMCA coursed down as LAD in the interventricular groove after giving rise to intermediate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

Pulmonary Diseases in July 2010. The nature of the pleural fluid was an exudate. Leucocyte count was 9.080/mm³, neutrophil count was 6.470/mm³, pH 7.51 and adenosine deaminase level was 10 U/L (<40 U/L). The albumin and lactate dehydrogenase levels of pleural fluid were 3.3 g/dL (3.5-5 g/dL), and 365 U/L (240-480 U/L), respectively. Left sided pleural fluid with mild pericardial fluid was detected on computerized tomography. The pleural fluid was thought to be due to parapneumonic effusion, anti-biotherapy and methylprednisolone were started. He was referred to our clinic in August 2010. He was investigated for possible diseases that lead to pleural effusion. His erythrocyte sedimentation rate (ESR) was 94 mm/h (0-20) and C-reactive protein (CRP) level was 24 mg/dL (0-0.5). Genetic analysis for Familial Mediterranean Fever mutation was negative. Antinuclear antibodies were negative. He did not have any sign of infection or systemic disease, although, he was still suffering from pleural and pericardial effusion. He used colchicine for 3 months but it did not make any difference. Amiodarone was discontinued because of its possible adverse effect. One month later there was no sign of pleural effusion on the chest X-ray, ESR and CRP levels were within normal ranges.

Amiodarone is a widely used anti-arrhythmic drug. Several types of pulmonary diseases such as chronic interstitial pneumonitis, organizing pneumonia, acute respiratory distress syndrome (ARDS), pulmonary fibrosis may occur in patients receiving amiodarone therapy (1). The incidence of pulmonary toxicity associated with amiodarone ranges from 1% to 10% (2). A high cumulative dose, duration of therapy, age, preexisting lung disease and surgery are defined as potential risk factors for developing pulmonary toxicity among patients treated with amiodarone-induced (1). It is speculated that amiodarone-induced pulmonary toxicity can be due to direct toxic injury to lung cells or indirect immunologic reaction (1). It has been reported that amiodarone-induced pleural effusion may be accompanied by the lung parenchymal involvement (3). However, one-third of the patients may have only pleural fluid accumulation (3). Corticosteroids have beneficial effects in treatment of amiodarone-induced pulmonary toxicity (1). In our case, pleuro-pericardial effusion was the only manifestation of pulmonary toxicity without a pre-existing lung parenchymal involvement. Therefore, in clinical practice, serositis findings in a patient receiving amiodarone can be ascribed to amiodarone after excluding other possible causes.

N. Şule Yaşar Bilge, Emel Gönüllü, Timuçin Kaşifoğlu, Cengiz Korkmaz
Division of Rheumatology, Department of Internal Medicine, Faculty of Medicine, Eskişehir Osmangazi University, Eskişehir- Turkey

References

1. Schwaiblmair M, Berghaus T, Haeckel T, Wagner T, von Scheit W. Amiodarone-induced pulmonary toxicity: an under-recognized and severe adverse effect? Clin Res Cardiol 2010; 99: 693-700. [CrossRef]
2. Jackevicius CA, Tom A, Essebag V, Eisenberg MJ, Rahme E, Tu JV, et al. Population-level incidence and risk factors for pulmonary toxicity associated with amiodarone. Am J Cardiol 2011; 108: 705-10. [CrossRef]
3. Uong V, Nugent K, Alalawi R, Raj R. Amiodarone-induced loculated pleural effusion: case report and review of the literature. Pharmacotherapy 2010; 30: 218. [CrossRef]

Address for Correspondence/Yazışma Adresi: Dr. Cengiz Korkmaz
Eskişehir Osmangazi Üniversitesi, Tıp Fakültesi, İç Hastalıkları Anabilim Dalı,
Romatoloji Bölümü, 26480 Eskişehir- Türkiye
Phone: +90 222 239 29 79 Fax: +90 222 239 37 74
E-mail: ckorkmaz@ogu.edu.tr

Available Online Date/Çevrimiçi Yayın Tarihi: 13.03.2012

©Telif Hakkı 2012 AVES Yayıncılık Ltd. Şti. - Makale metnine www.anakarder.com web sayfasından ulaşılabilir.

©Copyright 2012 by AVES Yayıncılık Ltd. - Available on-line at www.anakarder.com
doi:10.5152/akd.2012.081

Acute severe occlusion of the left main coronary artery following transcatheter aortic valve implantation



Transkateter aort kapak replasmanını izleyen akut ciddi sol ana koroner arter oklüzyonu

Transcatheter aortic valve implantation (TAVI) is an alternative therapy in patients with severe aortic stenosis (AS) and high surgical risk. Despite being less invasive than open-chest aortic valve replacement, TAVI remains to be associated with the potential for serious complications and one of them is coronary occlusion. Coronary occlusion during TAVI is a life-threatening complication that requires immediate diagnosis and treatment.

A 86-year old female patient with a history of hypertension, chronic obstructive pulmonary disease and coronary artery disease. Echocardiography demonstrated a severe calcified aortic stenosis with valve area 0.6 cm², mean transvalvular gradient 55 mmHg and left ventricular ejection fraction 55%. The distance between the annulus and the coronary artery ostia measured by multislice computed tomography (MSCT) scan was 10.2 mm. She had very high surgical risk (Logistic EuroSCORE=31.21%). She presented with dyspnea NYHA class III/IV, not responding to full medical treatment. A 23 mm Edwards Sapien prosthesis was successfully implanted percutaneously through the right femoral artery. After the valve deployment, patient developed ventricular fibrillation and hemodynamic collapse. Coronary angiography showed severe occlusion of the proximal left main coronary artery caused by the protrusion of a large calcium nodule of the native valve (Fig. 1, Video 1. See corresponding video/movie images at www.anakarder.com). The patient was treated with a 3.5x12 mm bare-metal stent, post-dilated with a noncompliant 4.0 mm balloon inflated at very high pressure. Final angiogram examination is shown in Figure 2 and Video 2 (See corresponding video/movie images at www.anakarder.com).



Figure 1 (Video 1). Coronary angiographic image of plaque shift resulting in severe left main coronary artery stenosis