Cukurova Medical Journal

Olgu Sunumu/ Case Report

Five Vessel Coronary Arter Bypass Graft Surgery in a Case with Familial Hypercholesterolemia

Familyal Hiperkolesterolemisi olan Bir Hastada Beş Damar Koroner Arter Bypass Cerrahisi

Süreyya Talay[1], Burçin Abud[2]

[1]Çanakkale State Hospital, Cardivascular Surgery. ÇANAKKALE

[2]Izmir Tepecik Research and Education Hospital.Cardivascular Surgery. İZMİR

Cukurova Medical Journal 2014;39(4):872-875.

ABSTRACT

We report a case of a rare and sypmtomatic familial hypercholesterolemia case with an end-point of coronary artery bypass surgery at the age of 16. Patient was evaluated at the emergency department with chest pain and discomfort. Physical examination were within normal limits. The electrocardiogram showed a normal sinus rhythm for 108/ min. Arterial blood pressure was 90/60 mmHg. Lungs and heart were clear to auscultation. Patient was under treatment with a prior diagnosis of familial hypercholesterolemia (FH) for one year by daily 40 mgs rosuvastatine. A coronary artery angiography was performed for chest pain. Multivessel coronary artery disease was diagnosed with a total occlusion of left anterior descending artery. Transthoracic echocardiography presented a left ventricular (LV) ejection fraction 50%, LV diameters 44/26 mm, aneurysm formation at interatrial septum and mild dyskinesia of anterior wall. Thus, a five vessel emergent coronary artery graft bypass surgery was performed at this early age. FH is with a severe elevation in total cholesterol (TC) and low density lipoprotein cholesterol (LDL) in an autosomal dominant characteristic disorder that approximately occurs in 1 per 500 persons by its heterozygous form. FH is most certainly associated with premature coronary artery disease (CAD) with catasthrophic early age results.

Key Words: Familial hypercholesterolemia, coronary artery bypass surgery, early age

ÖZET

Koroner arter bypass cerrahisi uygulanan familyal hiperkolesterolemili 16 yaşında bir hasta sunmaktayız. Acil servisimize göğüs ağrısı şikayeti ile başvuran hastanın fizik muayenesi olağandı.Elektrokardiyogram sinüs ritminde, kalp hızı dakikada 108 idi.Kan basıncı 90/60 mmHg olan hastanın kardiak ve solunumsal dinleme bulguları olağandı.Hasta 1 yıl önce familyal hiperkolesterolemi tanısı almış, bu sebeble günlük 40mg rosuvastatin kullanıyordu. Şikayetleri sebebiyle hastaya koroner anjiografi uygulandı ve sol ön inen koroner arterin (LAD) total oklüzyonunu içeren multidamar koroner arter hastalığı olduğu saptandı. Transtorasik ekokardiyografide ejeksiyon fraksiyonu %50, sol ventrikül çapları 26/44 mm ve ön duvarda orta diskinezi mevcuttu. Böylelikle bu genç hastaya beş damar koroner arter bypass cerrahisi uygulandı. Familyal hiperkolesterolemi düşük dansiteli lipoprotein (LDL) ve total kolesterolün ciddi oranda yükseldiği otosomal dominant bir hastalık olup, heterozigot formu 500 de 1 görülmektedir. Familyal hiperkolesterolomi genellikle erken yaşta mortaliteye yol olmaktadır, bununda sebebi erken yaşta meydana gelen koroner arter hastalığıdır.

Anahtar Kelimeler: Familyal hiperkolesterolomi, koroner arter bypass cerrahisi, erken yaş.

INTRODUCTION

We report a case of a rare and sypmtomatic familial hypercholesterolemia (FH) resulting with a multivessel coronary artery disease which was operated at the age of 16.

CASE REPORT

Patient administered to emergency department with chest pain and discomfort. He was receiving antihyperlipidemic medication of rosuvastatin 40 mgs, daily. Physical examination were within normal limits. The electrocardiogram showed a normal sinus rhythm for 108/ min. Arterial blood pressure was 90/60 mmHg. Lungs and heart were clear to auscultation. All arterial pulses at extremities and carotid regions were palpable and without any murmur to auscultation. Cardiac panel was normal by laboratory blood tests. His lipid profile was severely elevated one year ago by low-density lipoprotein cholesterol (LDL) 373.96 mg/dL, total cholesterol (TC) 436.23 mg/dL, triglyceride (TG) 339.71 mg/dL and high density lipoprotein (HDL) 32.97 mg/dL. He was receiving rosuvastatine 40 mgs a day by this initial diagnosis. Thus, at the point of administration, his lipid profile was recorded as LDL 135 mg/dL and 213 mg/dL TG.

We performed a coronary angiography due to chest pain. A multivessel coronary disease was detected at both left system (Figure 1) and right system (Figure 2). Left anterior descending coronary was totally occluded. Rest of the coronary tree was also presented severe segmental stenosis at all levels and vessels. Transthoracic echocardiography presented a left ventricular (LV) ejection fraction 50%, LV diameters 44/26 mm, aneurysm formation at interatrial septum and mild dyskinesia of anterior wall. Thus, symptomatic nature of patient's clinical features and angiographical findings, a coronary bypass surgery was mandatory.

Cardiovascular surgeons performed an emergent five vessel coronary artery bypass graft surgery by full arterial revascularization with left – right intermammarian artery, left radial artery and saphenous vein grafts. Patient was discharged at the postoperative day of ten without any complications.

DISCUSSION

FH is with a severe elevation in total cholesterol (TC) and low density lipoprotein cholesterol (LDL) in an autosomal dominant characteristic disorder that approximately occurs in 1 per 500 persons by its heterozygous form. On the other hand, homozygous FH prevalance is 1 in a million. Prevelance peak is observed in Ashkenazi Jews by 1 case in 67 persons[1,2].

FH is a result of a disorder of LDL receptor function which is due to a gene abnormality located on the short arm of chromosome 19. Homozygous forms present LDL levels over 600 mg/dL. Heterozygous forms may present LDL levels higher than 200 - 400 mg/dL. LDL levels over 330 mg/dL is an accepted threshold for diagnosis. TC levels are also elevated at the same time, generally more than 300 mg/dL. A detection of elevation of lipoprotein (a) (>30 mg/dL) presents a worsened prognosis among FH patients. However, a complete definition of FH diagnosis is determined by a gene analysis. Male to female ratio is approximately 1[3].

FH is most certainly associated with premature coronary artery disease (CAD) with catasthrophic early age results. Thus, each diagnosis of FH must be treated with an aggressive aspect with antihyperlipidemic medications to prevent the premature progression of coronary atherosclerosis. Clinical maifestations are reported to at the forth decade of life by cardiac morbidities and mortalities. Extracardiac morbidities are corneal arcus, xanthelasmas,

tendonitis, cardiac valve abnormalities, peripheral artery disease and xanthomas.

CAD complications are especially with co-morbidities such as HDL level less than 40 mg/dL, hypertension, smoking and positive family history (our patient's mother had a coronary surgery at the age of 38, father was hypertensive).

FH treatment must contain statins (HMG-CoA reductase inhibitors) at the maximum approved doses of rosuvastatin and atorvastatin. At inadequations, nonstatin cholesterol lowering medications such as bile acid sequestrants, nicotinic acid (niacin), gemfibrozil, fenofibrate and ezetimibe. Medication should aim 30-40% LDL reduction[4,5].

Any suspicion of cardiac and/or extracardiac vasculopathy in FH, a detailed examination of the cardiovascular system should be performed even by invasive tests such as coronary angiographies. Because of the severe nature of this genetic desease of FH, a catasthrophic myocardial infarction is always a part of the clinical feature. Furthermore, a diagnosis of CAD must be treated with a rapid onset of surgical intervention to prevent premature complications.

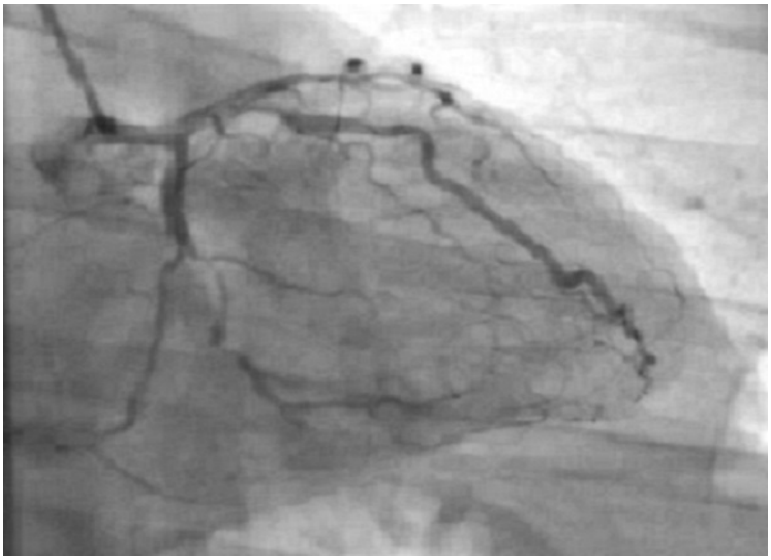

Figure 1. Preoperative angiography, left coronary system

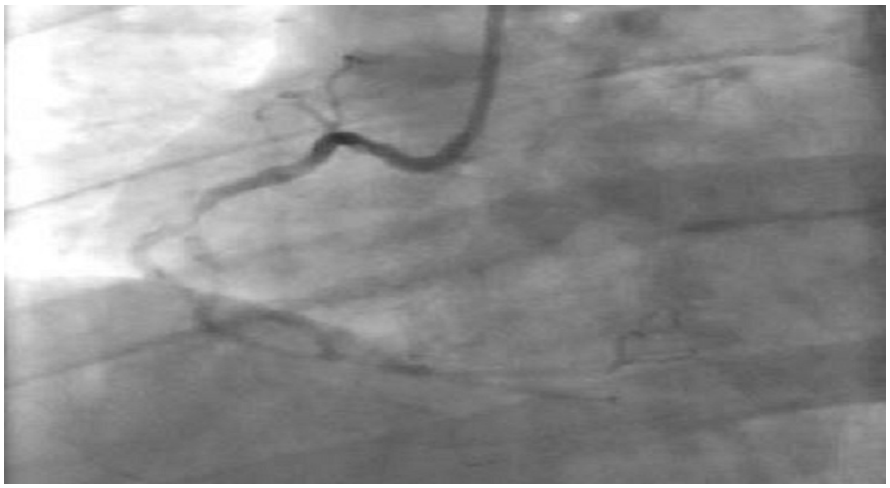

Figure 2. Preoperative angiography, right coronary system

REFERENCES

1. Ueda M. Familial hypercholesterolemia. Mol Genet Metab. 2005;86:423-6.
2. Wiesbauer F, Blessberger H, Azar D, et al. Familial-combined hyperlipidaemia in very young myocardial infarction survivors (<=40 years of age). Eur Heart J. 2009;30:1073–9.
3. Sibley C, Stone NJ. Familial hypercholesterolemia: a challenge of diagnosis and therapy. Cleve Clin J Med. 2006;73:57-64.
4. Gündüz H, Arınç H, Yolcu M, Kayardı M, et al. Familial Hypercholesterolaemia and Early Coronary Artery Disease: A Case Report and Review of the Literature. Turkiye Klinikleri J Med Sci. 2006;26:565-8.
5. Naukkarinen J, Ehnholm C, Peltonen L. Genetics of familial combined hyperlipidemia. Curr Opin Lipidol 2006;17:285–90.

Yazışma Adresi / Address for Correspondence:
Dr.Burçin Abud
İzmir Tepecik Research and Education Hospital
Cardivascular Surgery
İZMİR
E-mail: burcinabud@hotmail.com.

Geliş tarihi/received : 07.01.2014
Kabul tarihi/accepted:14.02.2014