

Hybrid treatment of supraaortic stenosis and coronary ostial stenosis in familial hypercholesterolemia

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Abstract

Familial hypercholesterolemia (FH) is an autosomal dominant inherited metabolic disorder characterized with high levels of low-density lipoprotein (LDL) plasma cholesterol. Increased plasma LDL-cholesterol levels cause cutaneous and tendinous xanthomas, supraaortic stenosis and coronary ostial stenosis in younger ages. Complications can be fatal if it is not diagnosed and treated in preliminary stages. We present a 22-year-old male patient with familial hypercholesterolemia and concomitant supraaortic stenosis and coronary artery disease treated with hybrid coronary revascularization approach.

Key words: Familial Hypercholesterolemia, Coronary Ostial Stenosis, Supraaortic Stenosis, Hybrid Approach

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Introduction

Familial hypercholesterolemia (FH) is a metabolic disorder caused by the mutations in the low density lipoprotein (LDL) cholesterol receptor gene.¹ The incidence of autosomal dominant inherited familial hypercholesterolemia is 1 in 1.000.000.^{2,3} Hypercholesterolemia not only affects the aortic root but also the aortic valve. Plaques formed by the accumulation of cholesterol particles lead supralvalvular aortic stenosis along with the atherosclerosis causing severe ostial stenosis of coronary arteries.⁴ Patients with this disorder might develop ischemic heart disease and myocardial infarction at early ages. In this case, we report a patient with autosomal dominant inherited familial hypercholesterolemia complicated with supralvalvular aortic stenosis and coronary artery disease who was surgically treated with hybrid approach.

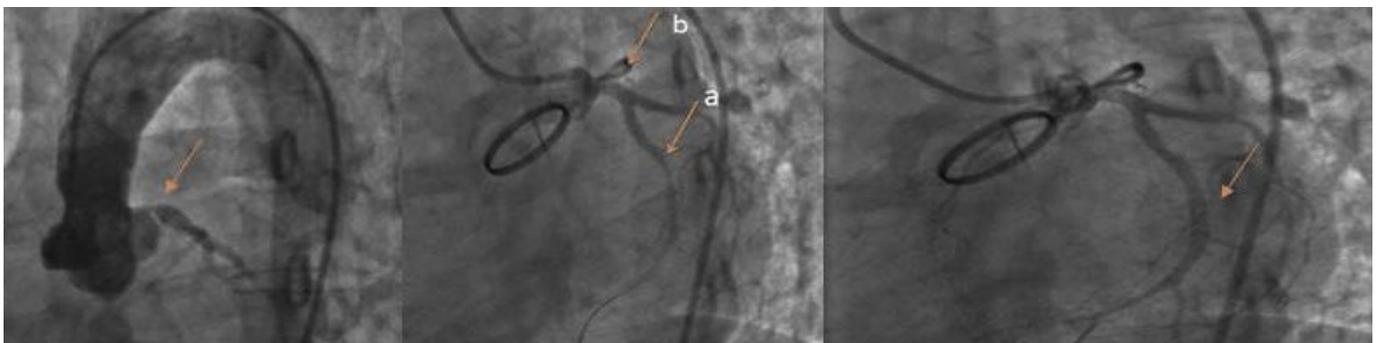
Case Report

A 22-year-old male patient was admitted to our department with symptoms of chest pain and dyspnea on exertion. He had been diagnosed with FH approximately 10 years ago. On admission he had xanthomas on his left elbow (**Figure 1**). He was a child of a consanguineous marriage. Although the use of HMG-CoA reductase inhibitor, laboratory studies of the patient, whose body surface area was 0.93 m², showed elevated serum total cholesterol, LDL cholesterol, high density lipoprotein (HDL) cholesterol and triglyceride levels which were 560 mg/dl, 450 mg/dl, 41 mg/dl and 146 mg/dl, respectively. Transthoracic echocardiography (TTE) showed combined supralvalvular aortic stenosis (peak/average gradient: 53/28 mmHg) and moderate aortic valve insufficiency. Angiographic examination revealed severe stenosis in the proximal left main coronary artery

Figure 1. Xanthomas on the left elbow.



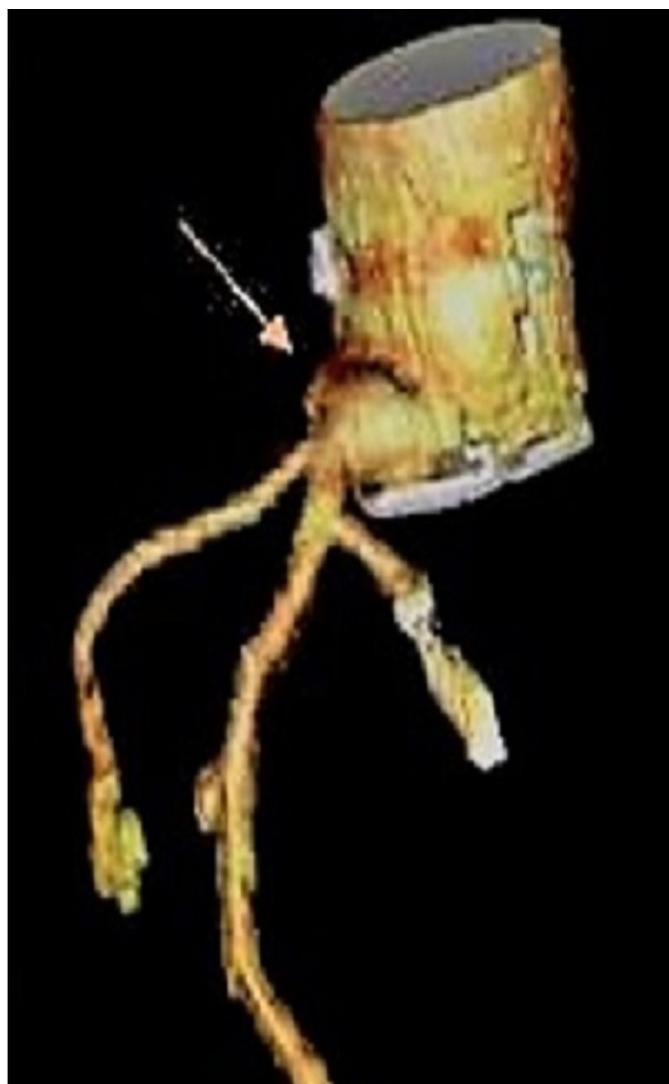
Figure 2. 90% ostial stenosis of LMCA, b. Stenosis (80%) in the mid portion of Cx artery.



(LMCA) and in the body of circumflex (Cx) artery as well as supra-ventricular aortic stenosis. **(Figure 2)**

Decision of surgical intervention was made according to these medical findings. Supracoronary ascending aorta and aortic valve were resected. Aortic valve replacement was performed with 21 mm St. Jude HP prosthesis. Then, ostial stenosis of LMCA was repaired with pericardial patchplasty and the patch was extended through the sinus of Valsalva. This surgical technique made the left sinus of Valsalva larger. **(Figure 3)** Subsequently, ascending aorta was replaced with a 26 mm Dacron graft. After two weeks of uneventful postoperative period, the cardiologist placed two metal stents to the Cx artery at cardiology unit. The patient was dis-

Figure 3. BT angiographic view of LMCA ostium after osteal patchplasty.



charged with anticoagulant, antiplatelet and statin therapies without any complications on the postoperative 16th day. The patient is in the follow-up period without any problem presently.

Discussion

Hyperlipidemia, premature atherosclerosis causing coronary artery disease, supra-ventricular aortic stenosis, large vessel atherosclerosis and giant xanthomas can be seen familial hypercholesterolemia. Atherosclerotic plaque of the aorta mostly develops in the abdominal segment in elderly patients, whereas premature atherosclerosis is often found in the ascending aorta and coronary ostia in patients with homozygous familial hypercholesterolemia.^{3,5} This condition leads to development of supra-ventricular aortic stenosis and ostial stenosis of the coronary arteries.

Treatment of homozygous FH consists of diet, medical therapy and apheresis. However, despite the medical treatment, majority of patients necessitate coronary revascularization and valve repair surgery. Coronary artery bypass grafting (CABG) with a full arterial grafting is a surgical option with longer patency rates in younger patients.⁶ In selected left main coronary artery lesions (noncalcified isolated lesions of the proximal or midportion LMCA) surgical patch angioplasty might be an alternative to CABG.

Surgical patchplasty restores patency of the narrowed coronary artery segment and increases physiological antegrade coronary artery blood flow. In LMCA lesions, if the coronary arteries are buried intramyocardially, the surgical patch angioplasty may be the only viable surgical treatment. If we use LMCA patchplasty in younger ages, classical CABG surgery and percutaneous stent implantation are available for distal coronary artery disease that might come out in future. Also, in case of a reoperation, the anatomy of the coronary graft is prevented from deterioration.

Autologous pericardium and saphenous vein are the most preferred patch materials with sufficient width and ease to suture. Malyshev et al. reported the rate of restenosis for autologous venous material and autologous pericardium were 3.4% and 4.9%, respectively.⁷ In our patient a young patient has osteal stenosis of LMCA and

stenosis of the Cx artery at the atrioventricular groove. In classic hybrid approach, minimal invasive LIMA to LAD bypass graft and percutaneous coronary intervention through to remaining lesions can be preferred.⁸ In our patient, we provided anatomical coronary ostial patency with patchplasty and hybrid coronary revascularization to the Cx artery in case of a need to a reoperation in the future.

Close follow-up is important in patients with familial hypercholesterolemia since ostial coronary artery

disease and ascending aorta disease with valvular involvement are likely to be seen at early ages of life. In LMCA lesions accompanying supra-ventricular aortic stenosis, ostial patchplasty combined with percutaneous coronary revascularization seems a better treatment option (especially in young patients) compared with conventional CABG, as there is no graft problem in redo surgery, the coronary anatomic plane is preserved and the higher capability of percutaneous interventions in concomitant distal coronary lesions.

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Disclosure and conflicts of interest:

The authors declare no conflict of interest.

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