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SHORT COMMUNICATION

Konno Ross procedure, coronary artery bypass graft and mitral valve replacement in a 12-year-old girl with homozygous familial hypercholesterolemia

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KEYWORDS

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Abstract *Background:* Familial hypercholesterolemia (FH) is a genetic disorder caused by a mutation of the gene for the low density lipoprotein receptor. This mutation can lead to elevated plasma cholesterol levels and subsequently to premature coronary artery disease. Management of patients with FH is complicated and surgery is accompanied by high risk, even in skillful hands. *Case presentation:* A 12-year-old female patient was referred to our department in January 2013 with chest pain and dyspnea. Her history showed that he had documented evidence of homozygous HF (HFH) since 2 years of age and that she underwent a Ross–Konno procedure for valvular aortic stenosis, 3 years ago. Electrocardiography showed ST depression in the inferolateral derivations and ST elevation in aVr. The echocardiography showed LV systolic dysfunction and important mitral regurgitation. Coronary angiography demonstrated stenosis in the distal part of the left main and severe three vessel coronary artery disease. The patient presented critical acute myocardial ischemia immediately after coronary angiography. She was referred for surgery. The left anterior descending artery was bypassed using saphenous vein and both right coronary artery and marginal artery using sequential saphenous vein. The mitral valve was replaced with mechanic prosthesis. The postoperative course was uneventful. She was prescribed atorvastatin accompanied by cholestyramine and diet modulation.

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Conclusion: HFH patients are at increased risk of developing coronary artery disease and also sudden death unless the condition is recognized and treated promptly. Surgery remains the most effective means of prolonging the life of these patients.

1. Introduction

Familial hypercholesterolemia (FH) is a genetic disorder caused by a mutation of the gene for the low-density lipoprotein receptor. This mutation can lead to elevated plasma cholesterol levels and subsequently to premature coronary artery disease. It may also lead to valvular and supravalvular aortic stenosis.

Management of patients with FH is complicated and surgery is accompanied by high risk, even in skillful hands.

Herein, we present a young patient with Homozygous FH (HFH) who presented with dyspnea and chest pain caused by coronary arteries stenosis and was treated with coronary artery bypass graft (CABG) surgery at the age of 12 years.

2. Case presentation

A 12-year-old female patient was referred to our department in January 2013 with chest pain and dyspnea. Her history showed that he had documented evidence of HFH since 2 years of age and that she had received lipid lowering drugs such as atorvastatin and cholestyramine. Besides, she underwent a Ross–Konno procedure for valvular aortic stenosis, 3 years ago.

On physical examination, she had multiple xanthomas (Fig. 1). Cardiac examination revealed sinus tachycardia of 120 bpm, blood pressure of 8/6 mmHg, crepitant rales and grade 3/6 systolic ejection murmur that was most prominent over the apex.

Electrocardiography showed ST depression in the inferolateral derivations and ST elevation in aVr.

The echocardiography showed LV systolic dysfunction and important ischemic mitral regurgitation. There were no signs of narrowing, insufficiency or calcification of the pulmonary autograft.

Troponin test was initially negative. Her lipid profile was as follows: total cholesterol, 5.27 g/l, LDL cholesterol, 4.75 g/l, HDL cholesterol, 0.29 g/l and TG, 1.19 g/l. The aortogram showed severe atherosclerosis of the ascending aorta and its branches (Fig. 2). Coronary angiography demonstrated stenosis in the distal part of the left main and severe three vessel coronary artery disease (Fig. 3).

The patient presented critical acute myocardial ischemia immediately after coronary angiography. She was referred for surgery. The left anterior descending artery was bypassed using saphenous vein and both right coronary artery and marginal artery using sequential saphenous vein. The mitral valve was replaced with mechanic prosthesis. The postoperative course was uneventful.

She was prescribed atorvastatin (40 mg/day) accompanied by Cholestyramine and diet modulation while waiting for the beginning of LDL-apheresis.

3. Discussion

FH is an autosomal dominant inherited disorder. Heterozygous FH occurs in approximately 1 in 500 people.[1] HFH is rare and occurs in approximately in 1 in 1 million persons

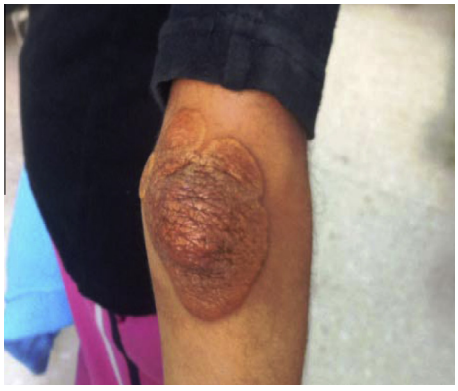

Figure 1 Xanthomas over patient's elbow.

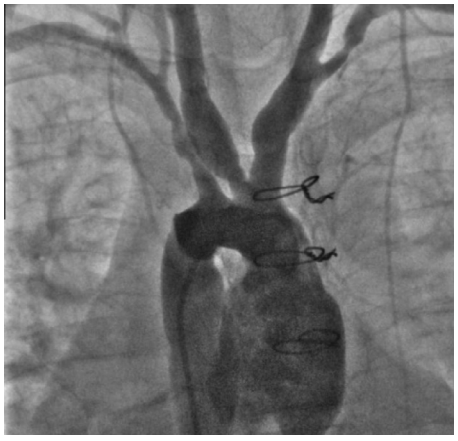

Figure 2 Aortogram showing stenosis in aorta branches.

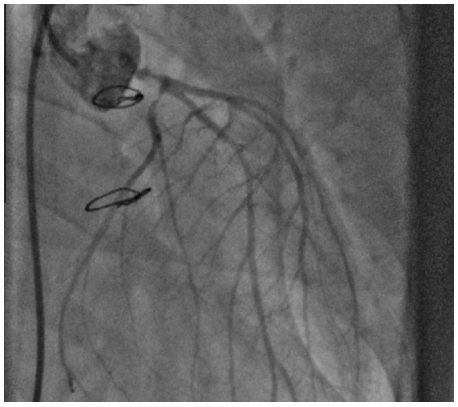

Figure 3 Angiography showing the left main stenosis.

worldwide and is a much more severe clinical disorder than heterozygous FH.

It is believed that total cholesterol level exceeding 9.68 mmol/l in children whose first degree relatives have hypercholesterolemia are highly suggestive for the disease.[2]

Premature coronary artery disease and atheromatous involvement of aortic valve and aortic root are the most serious complications of HFH which can result in clinical sequelae even in childhood.[3]

In our case, the atheromatous involvement of coronary arteries was present and severe atherosclerotic extent of the ascending aorta was also documented on the aortography.

Premature atherosclerosis might lead to cardiac ischemia and coronary revascularization is the last resort to alleviate the symptoms. This usually presents as angina but may also present with syncope, dyspnea or sudden death. The predilection for left main stem has been previously noted in HFH.[4]

Acute myocardial ischemia may occur after coronary angiography, especially in patients with HFH and coronary ostial stenosis. Our patient presented an acute myocardial ischemia immediately after the coronary angiography.

Currently recommended methods for coronary revascularization in HFH are coronary artery bypass grafting and stent implantation but pediatric experience remains scarce.

In fact, there is no available literature concerning the results of coronary angioplasty in children with HFH with the exception of few case reports.[5,6] Because of the rapid progression of coronary atherosclerosis in patients with HFH, a surgical strategy is preferred. Some researchers believe that arterial revascularization can be the best approach for the affected children[7,8] however, acceptable results from using venous grafts have been reported previously.[9] We used venous graft because angiography showed severe atherosclerosis of the ascending aorta and its branches. Concerning the ischemic mitral regurgitation, mitral valvoplasty was difficult in our case, so the surgeon opted for a valve replacement.

To the best of our knowledge, she is the youngest patient in the literature for whom a Ross–Konno procedure, a coronary artery bypass graft and mitral valve replacement have been done in childhood.

The management of HFH patients is a challenging job. Lipid lowering medications and lipid apheresis are two options to decrease serum cholesterol level; however, the best option to normalize the lipid profile might be liver transplantation.[10]

After the coronary artery bypass, we decided to pursue the statin therapy with lifelong antiaggregant acetylsalicylic acid treatment. Moreover, we started the therapy with LDL-apheresis since this approach is currently the treatment of choice for HFH and allows a significant lowering of cholesterol levels.

Stabilization or regression of coronary atherosclerosis has been reported with LDL apheresis in patients with HFH.[10]

4. Conclusion

HFH Patients are at increased risk of developing coronary artery disease and also sudden death unless the condition is recognized and treated promptly. Surgery remains the most effective means of prolonging the life of these patients. Long-term follow up and regular evaluation is recommended.

Conflict of interest

None.

References

1. Livia P, Claudia PO, Giovanni MP, et al. Autosomal recessive hypercholesterolemia and homozygous familial hypercholesterolemia: a phenotypic comparison. *Atherosclerosis* 2006;**188**:398–405.
2. Austin MA, Hutter CM, Zimmern RL, et al. Genetic causes of monogenic heterozygous familial hypercholesterolemia: a huge prevalence review. *Am J Epidemiol* 2004;**160**:407–20.
3. Haitas B, Baker SG, Meyer TE. Natural history and cardiac manifestations of homozygous familial hypercholesterolemia. *Q J Med* 1990;**76**:731–40.
4. Sprecher DL, Schaffer DJ, Kent KM, et al. Cardiovascular features of homozygous familial hypercholesterolaemia: analysis of 16 patients. *Am J Cardiol* 1984;**54**:20–30.
5. Mehan VK, Salzmann C, Pfammatter JP, Stocker FP, Meier B. Left main coronary angioplasty in a 10-year-old boy with homozygous familial hypercholesterolemia. *Cathet Cardiovasc Diagn* 1993;**29**:24–7.
6. Tissot C, Da Cruz E, Aggoun Y. Rescue left main coronary artery stenting for acute myocardial ischemia after coronary angiography in a 7-year-old girl with homozygous familial hypercholesterolemia. *Catheter Cardiovasc Interv* 2007;**69**:243–7.
7. Bhan A, Swain S, Juneja R, Saxena P, et al. Total arterial revascularization in a child with familial homozygous hypercholesterolemia. *J Thorac Cardiovasc Surg* 2005;**129**:932–4.
8. Goksel OS, Tireli E, El H, Oflaz H, et al. Coronary artery bypass grafting in a 12-year-old girl with familial hypercholesterolemia. *Acta Chir Belg* 2009;**109**:117–8.
9. Nemati MH. Aortic stenosis five years after coronary artery bypass grafting in a young patient with homozygous familial hypercholesterolemia. *J Heart Valve Dis* 2009;**18**(1):125–7.
10. Tatami R, Inoue N, Itoh H, et al. Regression of coronary atherosclerosis by combined LDL-apheresis and lipid-lowering drug therapy in patients with familial hypercholesterolemia: a multicenter study. The LARS investigators. *Atherosclerosis* 1992;**95**:1–13.