Full arterial revascularization with "no-touch" technique in a patient with familial homozygous hypercholesterolemia

Ailesel homozigot hiperkolesterolemili bir hastada "no-touch" teknikle yapılan full arteryel revaskülarizasyon

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Although familial homozygous hypercholesterolemia is a very rare pathology, it may be associated with severe cardiac disease. It causes high plasma levels of cholesterol with accelerated atherosclerosis resulting in the development of cutaneous xanthomas, premature coronary artery disease and aortic atherosclerosis. These patients may require early coronary artery bypass grafting operation. In this article, we describe a 22-year-old female patient with familial homozygous hypercholesterolemia who was treated successfully with a "no-touch" technique coronary artery bypass grafting operation because of severely atherosclerotic ascending aorta. The use of full arterial revascularization by using a "no-touch" technique prevents the risk of systemic embolization and also offers good long-term patency.

Key words: Atherosclerotic ascending aorta; coronary artery bypass grafting operation; familial homozygous hypercholesterolemia.

Ailesel homozigot hiperkolesterolemi oldukça nadir bir patoloji olmasına karşın ciddi kalp hastalıkları ile birlikte bulunabilir. Bu patoloji yüksek kolesterol plazma düzeyleri ve aterosklerozun hızlanmasına bağlı olarak ciltte ksantoma tarzı lezyonlar, erken koroner arter hastalığı ve aterosklerotik aort oluşmasına neden olmaktadır. Bu hastalara erken yaşta koroner arter bypass greftleme ameliyatları gerekebilmektedir. Bu yazıda çıkan aortta ciddi ateroskleroz nedeniyle "no-touch" yöntemi ile başarılı bir koroner arter bypass greftleme ameliyatı yapılan ailesel homozigot hiperkolesterolemili 22 yaşında kadın hasta sunuldu. "No-touch" tekniği ile yapılan full arteryel revaskülarizasyon sistemik embolizasyon riskini önlenmekte ve uzun dönem greft açıklığı sağlamaktadır.

Anahtar sözcükler: Aterosklerotik çıkan aort; koroner arter bypass greftleme ameliyatı; ailesel homozigot hiperkolesterolemi.

Familial hypercholesterolemia is a dominantly inherited disease which causes severe hypercholesterolemia because of increased levels of plasma low-density lipoprotein (LDL).[1] The disease process may be associated with premature coronary heart disease in addition to the development of cutaneous and tendon xanthoma.[1,2] We present here a 22-year-old female patient with familial homozygous hypercholesterolemia and severe coronary artery disease who underwent total arterial revascularization with aortic "no-touch" technique.

CASE REPORT

A 22-year-old female patient with the diagnosis of familial hypercholesterolemia and amyloidosis was admitted to our hospital. She had a strong familial history and her sibling died two years ago during an aortic valve replacement procedure. She was undergoing plasma LDL apheresis every 15-days and the laboratory examination just after the last apheresis revealed a total cholesterol level of 194 mg/dl, a LDL level of 40 mg/dl and a high-density lipoprotein (HDL) level of 14 mg/dl. On physical examination, there were an apical 2/6 pansystolic murmur and multiple tendinous xanthomas over knee and elbow joints (Fig. 1).

Her thallium scintigraphic evaluation was positive and subsequent cardiac catheterization revealed 80% stenosis of the left main coronary artery and 95% osteal stenosis of the right coronary artery (Fig. 2).

There were also non-stenotic atheromatous plaques at the right subclavian and left carotid arteries. Echocardiographic examination showed a mild degree of mitral and aortic insufficiency.

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On the basis of these findings, the patient was taken up for coronary artery bypass grafting. Following median sternotomy, gentle digital palpation of the ascending aorta also revealed circumferential calcification and bulky atheromatous plaque formations. Transoesophageal echocardiography demonstrated severe atherosclerotic disease of the ascending aorta (Fig. 3) and off-pump coronary artery bypass grafting operation using a "no-touch" technique was decided.

Bilateral internal thoracic arteries and left radial artery were then harvested, and following systemic heparinization, the left and right internal thoracic arteries were anastomozed to the left descending and right coronary arteries respectively by means of 7.0 polypropylene sutures on the beating heart (Estech® cardiac stabilizators). The left radial artery was then sutured to the second obtuse marginal artery with the same technique and the proximal anastomosis of the radial artery graft was placed into the left internal thoracic artery as a "Y-shaped" anastomosis. After the completion of the operation, the patient was taken to the intensive care unit and weaned off mechanical ventilation at the end of six hours. The postoperative course of the patient was uneventful and she was discharged on the 7th postoperative day. After the operation, the patient received antilipidemic therapy (cholestyramine and atorvastatin) and postoperative control angiography performed on the 6th postoperative month showed patent arterial grafts.

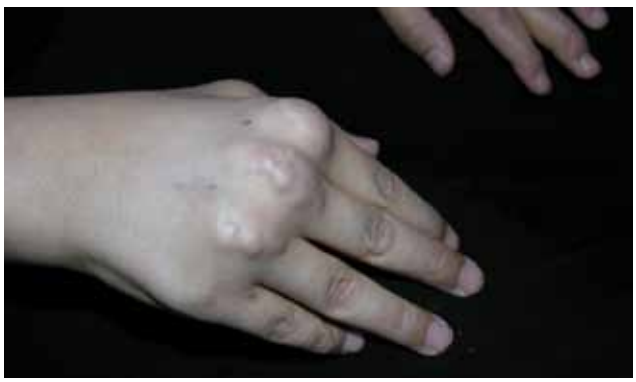

Fig. 1. The patient had multiple tendinous xanthomas over joints.

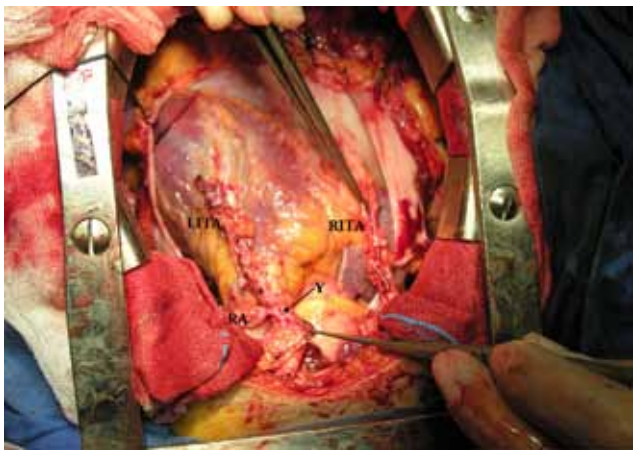

Fig. 2. Intraoperative view of triple bypass grafting. Left internal thoracic artery to left anterior descending, right internal thoracic artery to right coronary artery and radial artery to circumflex obtuse margin 1 artery. Radial artery was anastomozed proximally to left internal thoracic artery as Y graft.

DISCUSSION

Familial homozygous hypercholesterolemia is a rare pathology which may be associated with early atherosclerotic process affecting all body vessels including the ascending aorta and carotid arteries. In these patients, the major challenging issue complicating the intraoperative surgical strategy is the extent and degree of ascending aortic calcification. Since the manipulation of such a diseased aorta by aortic cannulation, cross-clamping and/or partial clamping may lead to intraoperative embolization, the use of a " no-touch" off-pump technique is strongly recommended while dealing with such patients.[3,4]

The benefits of internal thoracic and radial arteries have been well documented in several studies in terms of survival and freedom from symptoms.[5-8] Kawasuji et al.[1] stated that the internal thoracic arteries of these patients show no histologic differences from those of patients without familial hypercholesterolemia. Loop et al.[9] demonstrated that a better cardiac event-free survival rate may be obtained after internal thoracic artery grafting to the left anterior descending coronary artery. In view of these findings, we believe that a good long-term patency rate can be achieved with arterial grafts in patients with familial hypercholesterolemia. Additionally, since our patient was very young at the time of first operation, a full arterial revascularization

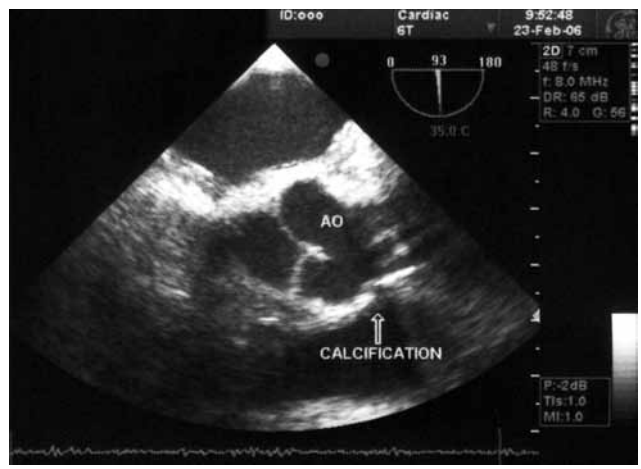

Fig. 3. Transoesophageal echocardiography demonstrated severe ascending aortic atherosclerosis.

procedure has been considered as the most acceptable approach to avoid the risk of an early redo operation.

As a conclusion, during a cardiac intervention in patients with familial hypercholesterolemia, severe atherosclerosis of the ascending aorta, carotid and vertebral arteries should be also considered. In case of severe plaque formation, the use of full arterial grafting by using a "no-touch" technique prevents the risk of systemic embolization and also offers a good long-term patency.

Declaration of conflicting interests

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