Off-pump coronary artery bypass grafting in a patient with neurofibromatosis I

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Abstract

Neurofibromatosis, otherwise known as von Recklinghausen disease, is a congenital hereditary disorder involving tissues of neuroectodermal or mesodermal origin. Involvement of the coronary arteries is, however, extremely rare. To the best of our knowledge, this represents the first report of off-pump coronary artery bypass grafting using the bilateral internal mammary arteries for coronary arterial vasculopathy in a patient with neurofibromatosis and of the longest reported follow-up of such a case after bypass surgery.

Keywords: Neurofibromatosis • Coronary disease • Off-pump coronary artery bypass grafting

INTRODUCTION

Neurofibromatosis, otherwise known as von Recklinghausen disease, is a congenital hereditary disorder involving tissues of neuroectodermal or mesodermal origin. Manifestations of this disease include pigmented skin lesions, cutaneous neurofibromas, tumours of the central/peripheral nervous gastrointestinal systems, skeletal abnormalities and vascular abnormalities. Involvement of the coronary arteries is, however, extremely rare. To the best of our knowledge, this represents the first report of off-pump coronary artery bypass grafting using the bilateral internal mammary arteries for coronary arterial vasculopathy in a patient with neurofibromatosis and of the longest reported follow-up of such a case after bypass surgery.

CASE REPORT

A 41-year old man, with a history of having been diagnosed as having neurofibromatosis type 1 several years ago, presented with a history of chest discomfort on effort since January 2008. He visited an outpatient clinic, where the presence of multiple café au lait skin macules and subcutaneous neurofibromata were recorded. He had no history of diabetes mellitus, hypertension, hyperlipidaemia or Kawasaki disease. Chest X-ray did not reveal any significant abnormalities, and the laboratory test results were unremarkable. An electrocardiogram showed the Q5 pattern and ST elevation in leads V1 and V2. Echocardiography also showed severe hypokinesis in the anterior septal wall and akinesis in the apex. The left ventricular ejection fraction was reduced to 46%. No valvular pathologies were noted. Coronary angiography revealed right coronary artery aneurysm with a mural thrombus, associated with 75% stenosis of the distal vessels, and 99% stenosis of the left anterior descending artery (LAD) (Fig. 1A and B). The remainder of the coronary artery vasculature appeared normal. As thallium scintigraphy confirmed viability of the myocardium in the anterior wall of the left ventricle, surgical revascularization was performed. Under general anaesthesia, the internal mammary arteries of both sides were harvested via a median sternotomy in a complete skeletonized fashion using a harmonic scalpel (Ethicon Endo-Surgery, Cincinnati, OH, USA). The left internal mammary artery was anastomosed to the LAD. Subsequently, the right internal mammary artery (RIMA), extended with the left radial artery, was anastomosed to the posterior descending artery. All vessels were very fragile, and special attention was required to avoid surgical injury. Operation time was 182 min, and no blood transfusion was required. The postoperative course was uneventful. At present, 5 years since the surgery, the patient remains well, and the follow-up coronary computed tomography reveals patent grafts and no additional aneurysmal changes in the native coronary artery or the internal mammary artery graft of either side (Fig. 2).

DISCUSSION

Neurofibromatosis is a congenital disorder of autosomal dominant inheritance that occurs at an incidence of ~1 in 3000 births [1]. There are two distinct neurocutaneous forms of the disease. The central type, or neurofibromatosis type 2, presents with multiple intracranial schwannomas, intracranial and spinal meningiomas, and ependymomas and spinal neurofibromas. The peripheral type, or neurofibromatosis type 1, also known as von Recklinghausen disease, is the more common (90%). It is caused by a mutation of a gene on the long arm of chromosome 17, which encodes a protein known as neurofibromin. It is characterized by dysplasia of the mesodermal and neuroectodermal tissues, and may involve
various organs of the body, with a potentially wide range of manifestations.

Brasfield et al. [2] reported that vascular anomalies occur in ~3.6% of these patients and are associated with a high mortality. The renal arteries are the most commonly affected vessels in patients with neurofibromatosis. However, involvement of the superior mesenteric, cerebral and thoracocervical arteries, including the subclavian vessels, have been reported. Vascular lesions include stenoses, occlusions, aneurysms, arterio-venous malformations and fistulae.

The mechanism of vasculopathy in neurofibromatosis remains unknown. Greene et al. [3] suggested two appearances of vasculopathy based on electron microscopic findings. First, large arteries may show intimal proliferation, which cause stenoses, medial thinning and fragmentation of elastic tissue, which may lead to aneurysm formation. These vessels may also be involved with neurofibromatous tissue. Secondly, small arteries show smooth-muscle aggregates within the intima, with resultant luminal narrowing.

Coronary artery involvement in neurofibromatosis is an extremely rare occurrence, although it is listed as one of the vascular manifestations of neurofibromatosis. Although actual prevalence of symptomatic or asymptomatic coronary artery disease among patients with neurofibromatosis I still remains unknown, average age of the onset of coronary artery disease was 41.7 years, and
57% of patients were <40 years old, from the 14 patients reported in the literature until now. Evrengul et al. [4] reported conventional single-vessel coronary artery bypass grafting using the left internal mammary artery in a 17-year old patient with neurofibromatosis. However, considering that malignant transformation and also tissue fragility is frequent in this disease, we strongly recommend off-pump coronary artery bypass grafting, which is beneficial for maintenance of the patients’ immune functions and avoidance of mechanical injury related to cardiopulmonary bypass.

In the present case, we utilized the internal mammary artery of both sides and the radial artery, because of the well-known long-term patency of these vessels and reports of good results. However, Conlon et al. [5] have reported the occurrence of spontaneous haemothorax caused by rupture of an internal mammary artery pseudoaneurysm in a patient with neurofibromatosis type 1. Therefore, we have to follow up the patient closely in the future to detect any possible aneurysmal change and stenosis or occlusion of the internal mammary arteries. Until now, 5 years since the surgery, the patient has not shown any adverse graft trouble. To the best of our knowledge, this is the first report of multiple lesions, including aneurysm, of the coronary arteries in a neurofibromatosis patient in whom off-pump total arterial coronary artery bypass grafting was performed, and at the 5-year follow-up, which is the longest reported till date in the literature, computed tomography demonstrated good patentcy without any new aneurysmal change, stenosis or occlusion in the coronary vasculature.

Conflict of interest: none declared.

REFERENCES


Figure 2: (A and B) Image of 3D computed tomography demonstrating right internal mammary artery (a) and left internal mammary artery (#).