Case report

Coronary artery dissection, combined aortic valve replacement and coronary bypass grafting in osteogenesis imperfecta

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Abstract

Osteogenesis imperfecta is an inherited connective tissue disorder. Aortic root dilation, aortic insufficiency and mitral valve prolapse are uncommon cardiovascular manifestations of osteogenesis imperfecta. Cardiac surgery in patients with osteogenesis imperfecta involves a high risk of complication rate. We report a case of coronary artery dissection induced by coronary angiogram in a patient with osteogenesis imperfecta and severe aortic regurgitation. In this case, the dissection of a coronary artery was not completely sealed by coronary stenting, and followed by successful combined aortic valve replacement and coronary artery bypass grafting on an emergency basis. © 2002 Elsevier Science B.V. All rights reserved.

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1. Introduction

Osteogenesis imperfecta is an inherited connective tissue disorder. Osteogenesis imperfecta is categorized in a group that includes Ehlers–Danlos syndrome, Marfan’s syndrome, Hurler’s syndrome and pseudoxanthoma elasticum. Osteogenesis imperfecta results from deletions, insertions or exon slice errors in the genes encoding type I collagen pro-
\(\alpha1\) and pro-
\(\alpha2\) chains. In most cases, the mutation is unknown and diagnosis is made by clinical assessments of symptoms, which include bone fragility, defective skeletal development, smaller stature, blue sclerae and hyperextensible ligaments. Cardiovascular abnormalities are infrequently documented in osteogenesis imperfecta. Aortic root dilatation, aortic insufficiency, and mitral valve prolapse are well known but are uncommon cardiovascular manifestations. Apparently rare defects in this disease are aortic dissections, significant coronary artery disease and coronary artery aneurysms. One case of spontaneous multivessel cervical artery dissection has been described [1], but no coronary artery dissection, either spontaneous or induced by catheter has been reported earlier in patients with osteogenesis imperfecta.

We present a case of dissection of the right coronary artery leading to successful combined coronary bypass grafting and aortic valve replacement on an emergency basis in a patient with osteogenesis imperfecta.

2. Case report

An 18-year-old man had a past history of recurrent long bone fractures, and osteogenesis imperfecta was diagnosed in the childhood. He had no current medication and his exercise tolerance was good (NYHA class I). On physical examination he was a man of short stature with blue sclerae. Auscultation revealed a diastolic grade 3/6 murmur at the right sternal border and an Austin Flint murmur. The blood pressure was 134/38 mmHg and the carotid pulse was steep with a high peak. The electrocardiogram (ECG) was unremarkable, except for voltage changes consistent with left ventricular hypertrophy. Transthoracic echocardiography showed left ventricular dilatation (LVEDD 76 mm), severe aortic regurgitation and mild mitral valve prolapse and regurgitation. Ejection fraction and aortic root diameter were normal. Cardiac catheterization was performed without any difficulties using 6F catheter. The left coronary angiogram was normal. After injection into right coronary artery (RCA), dissection of the entire proximal part of the RCA was observed (Fig. 1), and shortly after that the RCA occluded totally. This led to bradycardia and hypotension, and the patient felt chest pain and nausea. He was treated with medication and one stent implantation to the proximal

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part of the RCA. After stenting there was TIMI grade 1 flow in the vessel, but bradycardia and hypotension disappeared. Additional stenting was unsuccessful and the angiographic result remained suboptimal. Aortography showed severe grade 4/4 aortic regurgitation. The patient was operated immediately on an emergency basis. The aortic valve was replaced with a 23-mm mechanical aortic valve prosthesis and single saphenous vein aortocoronary bypass grafting to RCA was performed. His postoperative course was complicated by second degree atrioventricular block, which subsided by discontinuing the beta-blocker medication. The patient was discharged on the tenth postoperative day. At the 4-month follow-up, he felt well.

3. Discussion

Osteogenesis imperfecta results from structural defects in collagen, but the specific pathology of arteriopathy is unclear. According to Wheeler et al. [2], there is a marked decrease in the adventitial and intramural collagen of the intramyocardial arteries and great vessels in the lethal perinatal osteogenesis imperfecta (type II). The significant coronary artery disease and coronary aneurysms have been reported in patients with osteogenesis imperfecta [3]. We wanted to minimize unexpected difficulties during operation. Despite the young age of this patient we decided to perform an angiography. We assume that the dissection of the RCA was caused by the structural weakness of the coronary vessel wall, because there was no unusual manipulation of the diagnostic catheter. Stent may have been implanted into false lumen because additional stenting was unsuccessful. This is the first time when coronary artery dissection is reported in osteogenesis imperfecta.

Surgical procedures performed in osteogenesis imperfecta patients involve a higher than average risk of complications related to platelet dysfunction, friable tissue, impaired wound healing and musculoskeletal weakness and deformity [4]. Successful prosthetic aortic and mitral valve replacement has been reported in several patients with osteogenesis imperfecta. We could only find three case reports of successful coronary artery grafting and two case reports of combined aortic valve replacement and coronary bypass grafting in this disease entity in the English-language literature [3,5–8]. According to our knowledge all reported cases were elective operations. In our case, the dissection of the RCA led to the successful combined aortic valve replacement and coronary bypass grafting on an emergency basis. Despite the young age of this patient, saphenous vein was used in preference to the internal thoracic artery for revascularization because of sustained myocardial ischaemia during the operation.

Is it justified to perform an ascending aorta replacement, or even a Bentall operation, for avoiding further aortic dissection? It appears from all the reported cases that mortality after cardiac operations in patients with osteogenesis imperfecta is mainly due to friability of the tissue and bleeding. According to Houel et al. [9], early mortality is 7.7% with patient who undergoes a Bentall operation and 11% with patient who undergoes a separate replacement procedure of the ascending aorta and aortic valve. If the diameter of the aortic root and an ascending aorta is normal, the complication risk is too high in preventing operative management of the cardiovascular defects in this disease in urgent operation. Even in elective operation preventing management should not be performed routinely if the aortic diameter is normal or only mildly dilated.

In conclusion, the possibility of coronary artery dissection should be kept in mind while performing cardiac catheterization to patients with osteogenesis imperfecta and soft tip catheter should be used. Regardless of the high complication rate, urgent cardiac operations should be considered as a safe and effective treatment modality for patients with osteogenesis imperfecta.

References


