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A Loeys–Dietz patient with a transatlantic odyssey: repeated aortic root surgery ending with a huge left main coronary aneurysm

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Abstract

We present a patient who required several aortic root procedures. Finally, a huge aneurysm of the left main coronary artery required emergency surgery after a failed transcatheter procedure.

Keywords: Loeys–Dietz syndrome • Aortic root replacement • Left main coronary aneurysm • Graft infection

INTRODUCTION

Repeated aortic root surgery may be a challenging procedure. In the case of a prosthetic material infection, complete resection of the foreign material is mandatory in addition to adequate antibiotic treatment. Aortic homograft is a good option to control haemostasis and the infectious process. This Loeys–Dietz patient presented with a pseudoaneurysm of the left main coronary artery following multiple root procedures.

The 25-year old patient presented initially with aortic root aneurysm. Because of a very asymmetric pseudobicuspid aortic valve (Sievers type I, subcategory L–R) and the strong wish of the parents to avoid anticoagulation, he received a tissue-valved conduit in Bern in 2003 (Shelhigh Inc., NJ, USA—presently BioIntegral, Canada). Recovery was uneventful, but echocardiography at 6 months surprisingly demonstrated a limited dehiscence at the proximal anastomosis. Re-exploration showed that the suture line was not healed. Infection was excluded in the presence of normal clinical and laboratory findings and unsuspecting intraoperative inspection. A pericardial patch was used to reinforce the suture. Evolution was favourable.

In 2008, he presented with a degenerated tissue valve, requiring reoperation. The procedure was performed at Johns Hopkins University. The bio-conduit was resected, and a mechanical composite graft was implanted. The day after, the patient underwent repair of pectus excavatum. Recovery was uneventful.

Yearly echocardiographies and/or magnetic resonance imaging (MRI) were normal. In 2014, he developed fever and pneumonia was diagnosed. Blood cultures remained negative, but staphylococcus was revealed on polymerase chain reaction. Several weeks later, echocardiography and CT scan showed a partially thrombosed aneurysm of the left main coronary artery (Fig. 1). Positron emission tomography (PET) scan was not conclusive. The parents were against a fourth operative procedure, and asked several

opinion-leaders for a transcatheter technique. A covered stent was recommended to exclude the aneurysm. However, stable positioning of the stent between the ostium and the distal left main coronary artery was not successful, and obstruction of the left main coronary artery occurred requiring resuscitation. Percutaneous extracorporeal membrane oxygenation (ECMO) was installed. The patient was immediately moved to the operation theatre. Echocardiography showed a left ventricular ejection fraction (LV-EF) of 10–15%.

Re-sternotomy was performed, and ECMO was switched to cardiopulmonary bypass through central cannulation. Intraoperative inspection showed thrombotic material inside the graft. To expose the aneurysm, pulmonary artery was transected above the valve. Thrombotic material was removed. Continuity of the left main coronary artery was restored with a short vein graft. Aortic homograft was implanted in the mini-root technique, and pulmonary artery was re-anastomosed during reperfusion. ECMO support was necessary during 48 h. The patient was discharged after 5 weeks with still compromised LV function (EF 25–30%).

Antibiotics were continued for 6 weeks, although intraoperative cultures remained negative. Eighteen months postoperatively, the LV-EF has stabilized at 40%. The patient is doing well, and the findings are normal (Fig. 2).

COMMENT

Repeated aortic root surgery is a surgical challenge [1]. Aneurysm of the left main coronary artery is infrequent, but may occur in patients with connective tissue disease or as a result of infection.

This patient's history presents several critical decision-making points:

- (i) Initial biological composite graft was probably a wrong decision. A mechanical composite graft would have been the best

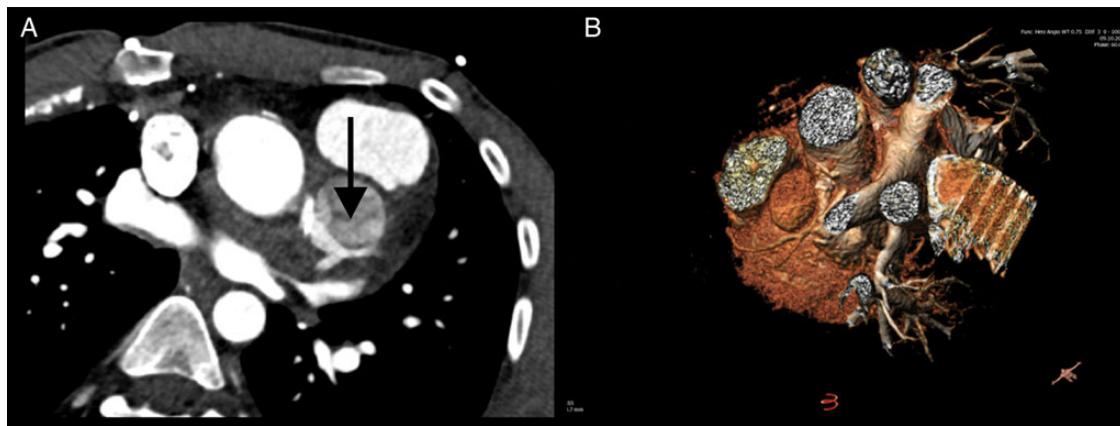


Figure 1: A (Pseudo)-aneurysm of the left main coronary artery on conventional CT scan (A) and 3D reconstruction (B).



Figure 2: Postoperative 3D reconstruction of the aortic homograft with separate interposition of two short vein grafts for the left and right coronary arteries.

long-term option since in 2003, a valve-sparing procedure was not an established standard in young patients with connective tissue disease and pseudobicuspid valve. The parents were strictly against anticoagulation.

- (ii) Dehiscence of the suture observed early postoperatively is a problem that we and others have described following implantation of this type of biological conduit [2, 3].

- (iii) Aetiology of the coronary aneurysm was uncertain. PET scan was not conclusive, and intraoperative cultures remained negative.
- (iv) Transcatheter attempt to exclude the left main coronary artery aneurysm was not a good decision. Sudden coronary obstruction led to resuscitation, and required immediate ECMO support.
- (v) Transection of the main pulmonary artery greatly facilitated exposure of the left main coronary artery.

Surgery was challenging because re-sternotomy was performed under resuscitation and after repaired pectus excavatum. Aortic homograft was found the best option to control intraoperative haemostasis [1]. Re-attachment of the coronary arteries required short vein graft interposition to avoid tension and exclude the aneurysm.

We conclude that indication for surgery should never be weakened because transcatheter intervention seems theoretically possible. In this case, catheter intervention turned the repair into an emergency situation and compromised significantly the myocardial function. In addition, decision-making was challenged by the opinion of non-expert relatives instead of a decision based on experience and expertise on technical adequacy.

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