



Editorial

Percutaneous Implantation of the self-expanding valve Prosthesis a patient with homozygous familial hypercholesterolemia severe aortic stenosis and porcelain aorta



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ABSTRACT

Transcatheter aortic valve implantation (TAVI) has shown favorable outcomes in patients with severe symptomatic aortic valve stenosis who are at high surgical risk or inappropriate for open heart surgery. However, concerns exist over treating patients who have porcelain aorta and familial hypercholesterolemia, due to the potential complications of aortic root and aortic annulus. In this case report, we present a patient with familial hypercholesterolemia, symptomatic severe aortic stenosis, previous coronary artery bypass grafting and porcelain aorta, who was successfully treated with TAVI using a CoreValve.

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1. Introduction

Familial hypercholesterolemia (FH) is characterized early appearance of cutaneous and tendon xanthomas, premature cardiovascular disease and atheromatous involvement of the aortic valve and root “porcelain aorta”. Porcelain aorta represents excessive calcification of the thoracic ascending aorta and is a challenging substrate for cardiac surgeons because aortic cross-clamping and aortotomy may cause excessive aortic injury and/or release of thromboembolic material that may cause periprocedural stroke. Aortic valve replacement in patients with a porcelain aorta may mandate advanced surgical techniques including replacement of the ascending aorta under deep hypothermic circulatory arrest, endarterectomy, or balloon assisted endovascular clamping to minimize manipulation of the heavily calcified aorta. Transcatheter aortic valve implantation (TAVI) represents a highly attractive “no-touch” alternative for patients with severe aortic stenosis and porcelain aorta.

2. Case report

A 59-year-old woman was admitted for frequent chest discomfort and dyspnea. The patient had been diagnosed with familial hypercholesterolemia and hypertension 13 years ago. She had a coronary artery bypass

grafting surgery for two vessels (Lad-Lima, Safen-Rca-Om) in 2003. On admission, she had marked tendon xanthomas of the upper and lower extremities and xanthelasma corners of her eyelids (Fig. 1). Although she was being treated with 80 mg of atorvastatin and 10 mg of ezetimibe, her total cholesterol level was 454 mg/dL, her low density lipoprotein (LDL) level was 314 mg/dL, her high density lipoprotein (HDL) level was 70 mg/dL, and her triglyceride level was 177 mg/dL. Cardiac examination revealed sinus tachycardia of 105 bpm, a blood pressure of 110/75 mm Hg, and a grade 3/6 systolic ejection murmur that was most prominent over the second intercostal space on the right sternal border. Electrocardiography showed left ventricular hypertrophy (Skolow-Lyon index) and ST depressions in the inferolateral derivations. Transthoracic echocardiography revealed normal left ventricular systolic function, hypertrophy of the left ventricle, and calcific aortic stenosis with a peak/mean gradient of 109/70 mm Hg and aortic valve area (AVA) of 0.4 cm². Coronary angiography demonstrated a 90% narrowing in the RCA-OM sequential saphenous vein graft and aortography showed porcelain aorta (Fig. 1). According to the statement of the European Association for Cardio-Thoracic Surgery (EACTS) and the European Society of Cardiology (ESC) the heart team’s role is to confirm the severity of aortic stenosis [1], a transcatheter aortic valve implantation was preferred due to the presence of a porcelain ascending aorta (logistic EuroSCORE 23%, Society of Thoracic Surgeons mortality score 5%).

As a routine pre-procedural approach, besides standard transthoracic echocardiogram, transesophageal echocardiogram (TEE) and multislice computed tomography (MSCT) has also been performed

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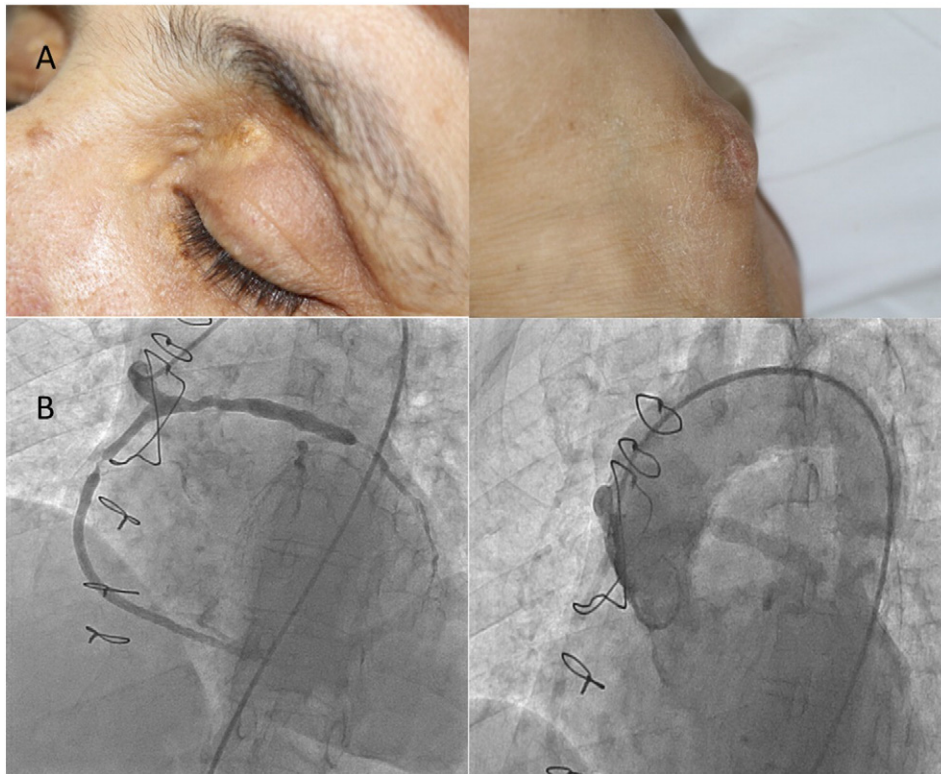


Fig. 1. A: Showed marked tendon xanthomas of the upper extremitie and xanthelasma corners of her eyelids B: Coronary and aortic angiogram showing narrow Safen-RCA-OM sequential greft and porcelain aorta.

before TAVI, in order to carefully assess (i) aortic root diameters, (ii) peripheral arterial access and (iii) burden of atherosclerosis. Multi-slice computed tomography scan revealed porcelain aorta, heavily

calcified atherosclerotic involvement of entire aorta, iliac and femoral arteries, but left common femoral artery was considered appropriate vascular access. TEE showed diffuse protruding into lumen

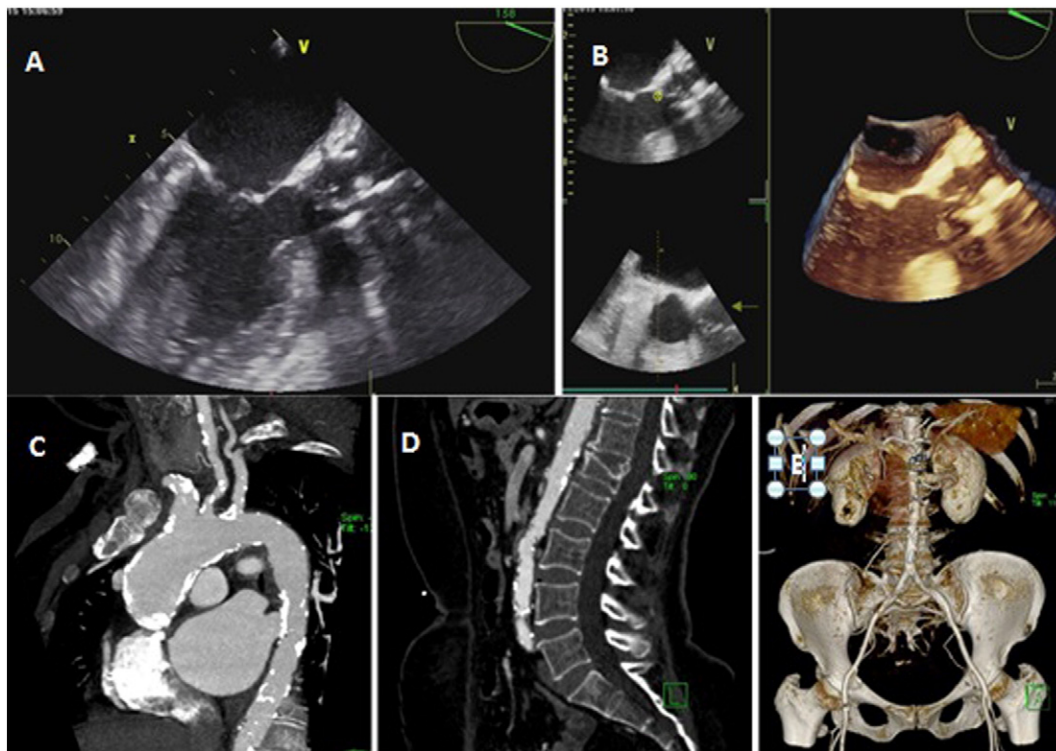


Fig. 2. A, B: Transesophageal echocardiography (TEE) and 3D reconstruction view showing diffuse protruding plaques in the ascending aorta and sinuses valsalva. C, D, E: Multislice cardiac computed tomography and 3D multislice cardiac computed tomography reconstruction showing porcelain aorta, heavily calcified atherosclerotic involvement of entire aorta, iliac and femoral arteries.

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