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Multi-Modality Guided Trans-Apical Closure of Recurrent Mitral Para-Valvular Regurgitation After Failed Surgical Management in a Patient with Osteogenesis Imperfecta

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We report a case of a 54 year-old man with osteogenesis imperfecta who developed severe para-valvular mitral regurgitation after a second heart operation to correct the same problem. The large para-valvular leak was successfully closed with an Amplatzer Vascular Plug III delivered from the apical approach.

Keywords Paravalular leak • Device closure • Transcatheter • Mitral • Trans-apical

Introduction

Para-valvular regurgitation as a result of partial valve dehiscence is a significant complication of prosthetic heart valves, most commonly affecting the mitral valve [1,2]. An important predisposing factor is abnormal tissue friability [3]. We report a case of a 54 year-old man with osteogenesis imperfecta who developed severe para-valvular mitral regurgitation after a second heart operation to correct the same problem. The para-valvular leak was successfully closed with an Amplatzer Vascular Plug III delivered from the apical approach.

Case Report

A 54 year-old man with osteogenesis imperfecta diagnosed at the age of 10 years underwent prosthetic aortic and mitral valve replacement in 1999 for severe incompetence of both valves. In 2001 he underwent a re-do mitral valve replacement (St. Jude 25 mm valve) for a significant lateral para-valvular leak associated with haemolysis. Subsequent trans-thoracic echocardiograms (TTE) after February 2002 showed apparently mild mitral valve regurgitation, the nature and extent of which could not be accurately determined because of acoustic valve shadowing.

In February 2013 he became progressively short of breath (NYHA Class 3) and anaemic (haemoglobin 101gm/l) with evidence of haemolysis. A trans-oesophageal echocardiogram (TEE) demonstrated left ventricular systolic function at the lower limit of normal, mitral incompetence the severity of which could not be accurately determined and an estimated pulmonary artery pressure of 60 mmHg compared to 30 mmHg on the previous TTE of 2008. A 3D (TEE) demonstrated moderate to severe mitral regurgitation due to a large crescentic shaped lateral para-valvular defect (dimensions

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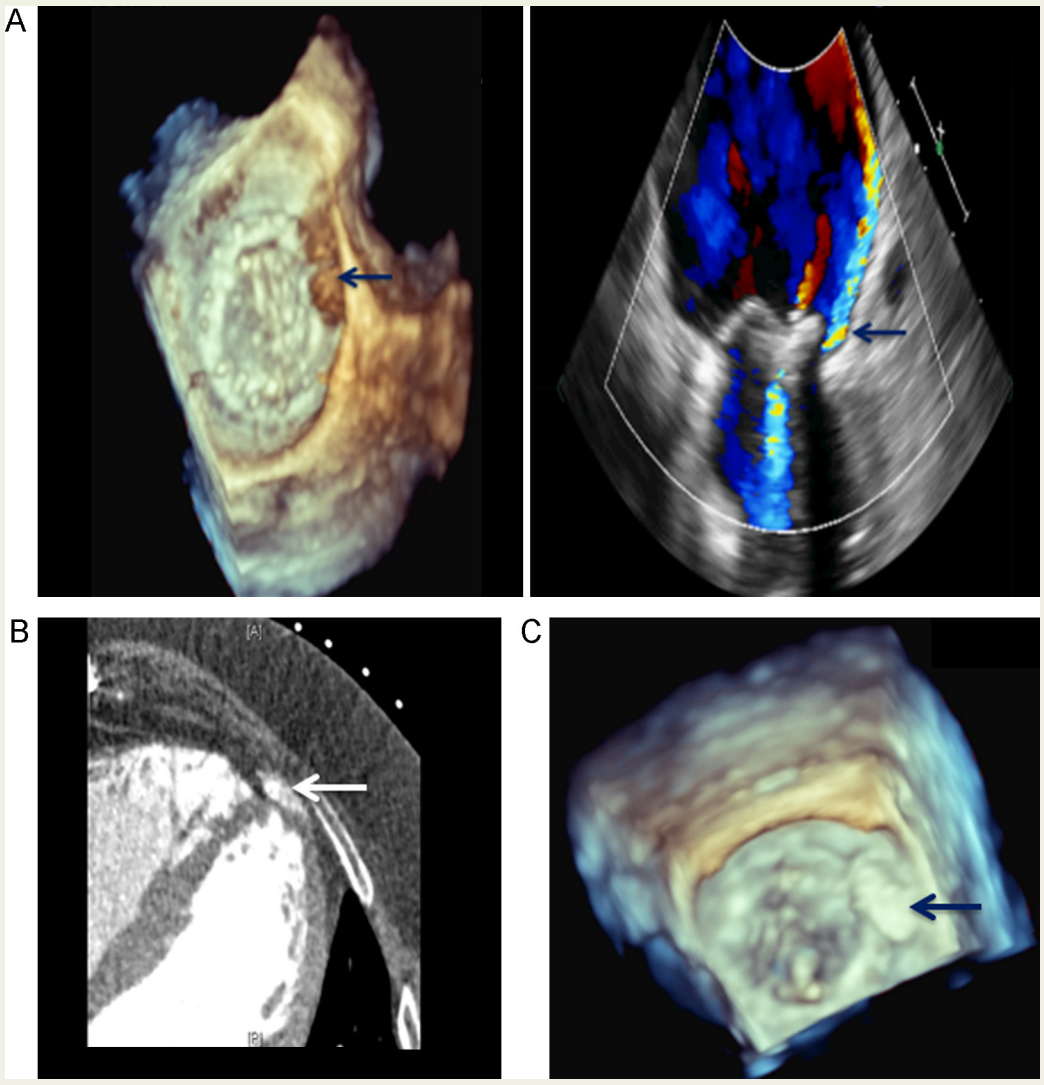

Figure 1 A 2D and 3D trans-oesophageal echocardiogram (TEE) showing moderate to severe mitral regurgitation due to a large crescentic shaped lateral para-valvular defect (arrow).
Figure 1B A cardiac computed tomogram (CT) (arrow) showing localised area of pericardial calcification in the apical region, because of which a surgical apical approach was favoured.
Figure 1C 3D trans-oesophageal echocardiogram showing the Amplatzer Vascular Plug III device (St Jude Medical) (arrow) well seated across the para-valvular defect with no residual para-valvular regurgitation.

15 mm x 4 mm) (Figure 1A) which was in a similar position to the previous para-valvular leak. A cardiac computed tomogram (CT) done to guide trans-apical access revealed a localised area of pericardial calcification in the apical region (Figure 1B).

A decision was made to attempt device closure of the defect. The procedure was done under general anaesthesia in the cardiac catheterisation laboratory with intra-procedural 3D TEE and fluoroscopy guidance. The left ventricular apex was exposed through a small left antero-lateral thoracotomy. A purse string suture with Teflon pledgets was placed to control bleeding. The left ventricular apex was punctured under direct vision and a short 8 French sheath inserted. A 6 French Amplatzer left coronary guiding catheter was inserted through the sheath and used to facilitate the steering of an angled hydrophilic guide wire across the defect into the left atrium. The 8 French Sheath was then exchanged for a longer 8 French hydrophilic sheath which was inserted into the left atrium over the guide wire. A 14 mm x 5 mm Amplatzer Vascular Plug III (St Jude Medical) was chosen, as it was the closest in size to the defect and the largest available device. It was then delivered through the sheath and opened across the defect using the 3D TTE to guide correct orientation of the device. Following delivery of the device the 3D TEE confirmed complete abolition of the paravalvular regurgitation and no impingement of the device on the prosthetic