defect and relieve the airways obstruction at the same time. The Lecompte maneuver with or without PA plication or competent right ventricular outflow tract conduit implantation represents the treatment for relief of airway compression due to dilated pulmonary arteries [4]. An alternative approach could have been to perform the Lecompte maneuver and place a limiting left ventricle to PA conduit, but that would have sacrificed the pulmonary (neoaortic) valve and required a ventriculotomy. Although the double switch is a complex procedure, we have previously shown that it can be performed in infants and in high-risk situations such as this with an actuarial survival at 10 years of 83.9% [5]. Similar results for double switch operation are reported from other centers, and it can be considered an optimal procedure to treat symptomatic ccTGA [6, 7]. This case report describes a very rare association of anomalies for ccTGA and shows the benefit of the Lecompte maneuver in bringing aneurysmal PAs away from the major airways in infants with extrinsic compression.

References


Fatty Infiltration of an Aortic Valve
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Adipose tissue is a normal constituent of the heart, but not a normal anatomic finding of cardiac valves. Fatty infiltration of the aortic valve is rare, with unknown significance on valve function. We report a case of fatty infiltration and replacement of the spongiosa layer in an incompetent aortic valve. The mechanism of fat infiltration is unknown, but may be explained by differentiation of preexisting valve interstitial cells secondary to valvular injury.

Fig 3. Chest computed tomography scan. (A) Black arrows indicate compression of the proximal right bronchus and proximal left bronchus by enlarged right pulmonary artery (*). (B) Posteroanterior view: white arrow points to the complete occlusion of the left bronchus and partial occlusion of right bronchus (three-dimensional reconstruction).

Accepted for publication Dec 31, 2012.
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Published by Elsevier Inc
http://dx.doi.org/10.1016/j.athoracsur.2012.12.061
heart disease, hypertension, hypercholesterolemia, and atrial fibrillation, for which she was anticoagulated.

On examination, the patient was not overweight, with a body weight of 60 kg. Her blood pressure was 170/70 mm Hg and irregular heart rhythm with rate 120 beats per minute. Signs of pulmonary edema and heart failure were present with an elevated jugular venous pressure, crepitations, and wheeze throughout both lung fields and peripheral edema.

Laboratory investigation revealed no abnormality. In particular, her lipid profile was normal, with cholesterol, triglycerides, low-density lipoprotein, and high-density lipoprotein being 3.1, 0.98, 1.46, and 1.21 mmol/L, respectively.

Transthoracic echocardiography demonstrated mild to moderately impaired left ventricular function (ejection fraction 0.44) with moderate to severe regurgitation of a tricuspid aortic valve. The distal anterior and inferior septal walls were akinetic, probably related to an old myocardial infarction. The aortic valve cusps were mildly echodense with normal excursion. There was mild mitral regurgitation and the aortic root was dilated at 4.9 cm.

Coronary angiography revealed an occluded non-dominant right coronary artery, with a patent stent in the left anterior descending artery. Aortography revealed a dilated aortic root and ascending aorta with severe aortic regurgitation.

After medical optimization, isolated aortic valve replacement was performed utilizing a 23-mm bioprosthetic valve (Carpentier-Edwards PERIMOUNT Magna Ease; Edwards Lifesciences Corp, Irvine, CA). The native aortic valve was sent for histopathologic analysis. Macroscopically, the cusps (measuring 20 to 30 mm) showed mild calcification with no other abnormality. Microscopically, the valve leaflets showed marked myxoid degeneration and fatty infiltration of the spongiosa extending to the annulus (Fig 1A). Myxoid degeneration was confirmed by extensive positivity of the stroma on alcian blue stain. Mature adipocytes were present and confirmed by positive S-100 on immunohistochemical studies (Fig 1B). No microscopic evidence of vegetation or valvulitis was present. The patient recovered well on the ward and discharged home on postoperative day 14.

Comment

Adipose tissue is a normal constituent of the heart, usually located in the epicardium. Fat infiltration into the myocardium (cor adiposum), interatrial septum, and lipomatous metaplasia in ischemic scar tissue of the myocardium have all been described [2, 3]. Fat is not a normal anatomic finding in cardiac valves. The aortic valve cusp is a laminate structure consisting of 3 primary layers, each defined by its own characteristic matrix composition and role in valvular function. The fibrosa, on the outflow side of the aortic valve, consists of circumferentially oriented collagen fibers that provide tensile strength. The ventricularis is a dense sheet of elastic fibers on the inflow side of the valve that is compliant and provides elasticity and preload to the leaflet. In-between the fibrosa and ventricularis is the spongiosa, where glycosaminoglycans and proteoglycans are believed to confer flexibility, dampen vibrations from closing, and resist delamination [4]. Any change in the constituents of any layer may adversely affect the function of the cusp.

To our knowledge, only 1 other publication in the English literature reports a finding of fatty infiltration of the aortic valve [1]. In this case report, a normally functioning aortic valve was replaced due to a complication of mitral valve repair. Prior to surgery, a tranesophageal echocardiogram revealed a mass in the right coronary cusp. Macroscopically the cusp contained yellow...
fatty foci. Microscopically, aggregates of infiltrating adipocytes were found only in the ventricularis of the right aortic cusp and did not extend to the aortic valve annulus.

Our report is the first to show near complete replacement of the spongiosa of all 3 aortic cusps with mature adipocytes extending to the aortic annulus. The significance of this finding on valve function is unknown. Although our patient had severe aortic regurgitation, it is questionable as to whether fatty infiltration into the aortic valve was causing the functional abnormality as there was significant dilatation of the aortic root and ascending aorta. Due to the patient’s age and comorbidities, replacement of the aortic root and ascending aorta was not performed.

Fatty infiltration could be explained by differentiation of valve interstitial cells (VICs) secondary to valvular injury from functional regurgitation. The VICs are a heterogeneous population of fibroblasts, with a small population of myofibroblasts and smooth muscle cells. In diseased states, this population of cells includes a greater proportion of myofibroblasts [5]. In porcine valves, this pathologic VIC phenotype likely originates from a subpopulation of mesenchymal progenitor cells with differentiation potential to myofibrogenic, osteogenic, chondrogenic, and adipogenic lineages [6]. Although similar mesenchymal progenitor cells are thought to exist in humans, they have not yet been identified [7]. It is perhaps this phenotypic plasticity of pathologic VIC that has led to adipocyte infiltration into the spongiosa of our reported aortic valve.

References


Early Bioprosthesis Mitral Valve Failure Due to Fusion of the Cusps

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Modern extracorporeal membrane oxygenation support has revolutionized the survival outcome of critically ill patients with refractory cardiac failure. Mitral valve replacement in patients treated with extracorporeal membrane oxygenation poses a surgical challenge regarding the choice of appropriate prosthesis. We present two cases of mitral valve bioprosthetic failure within days to weeks after valve replacement. Both patients were supported with extracorporeal membrane oxygenation postoperatively, and the bioprosthetic failure was caused by fusion of the cusps. This mechanism of mitral valve bioprosthesis failure in combination with extracorporeal membrane oxygenation treatment has not previously been described.


M itral valve replacement (MVR) is the treatment of choice when mitral valve plasticity is not feasible. The valve can be replaced with either a biological valve prosthesis or a mechanical valve prosthesis. Mechanical valves require a mandatory life-long use of anticoagulant therapy, whereas bioprosthetic valves have the disadvantage of a limited durability. Extracorporeal membrane oxygenation (ECMO) support is increasingly used in heart failure, before and after mitral valve replacement (MVR) [1]. Patients undergoing MVR with postoperative need for ECMO support pose a surgical challenge regarding the choice of appropriate prosthesis. The heparin-coated ECMO systems allow a reduced systemic anticoagulation, and it can even be entirely omitted in case of severe bleeding. Therefore, in patients treated with ECMO or when there is a high risk of requiring ECMO support perioperatively or postoperatively, the option of choosing a bioprosthesis could be considered a more attractive choice. We present, to our knowledge for the first time, two cases of mitral bioprosthetic valve failure due to fusion of the cusps occurring within days to weeks after implantation of the prosthesis in patients treated with ECMO postoperatively.

Case Reports

Patient 1

A 25-year-old man presented to the emergency department with a history of chest pain over a few hours. An ST-segment elevation myocardial infarction was diagnosed, and a drug-eluting stent was inserted in the left anterior descending artery. However, severe heart failure could not be avoided, and an Impella 5.0 device (Abiomed, Danvers, MA) was implanted. It was later on replaced by an intraaortic balloon pump and thereafter, venoarterial ECMO treatment (CentriMag; Levitronix, Waltham, MA; Bioline coated tubing, Quadrox

Accepted for publication Nov 16, 2012.

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