Untreated tetralogy of Fallot in an adult patient complicated by acute aortic valve endocarditis

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Abstract

Acute endocarditis complicated by aortic valve regurgitation is an uncommon finding in adults with surgical untreated tetralogy of Fallot (TOF). The hemodynamic consequences for both, the right and left ventricles, are significant. However, the right ventricle may be in a disastrous situation, since a compromised right ventricle from longstanding pressure overload may not tolerate acute volume overload. Here we report a 28-year-old African adult patient with TOF and acute severe aortic valve regurgitation due to bacterial endocarditis with preoperative low cardiac output syndrome. After aortotomy a large abscess cavity underneath the left and non-coronary sinus and a TOF typical perimembranous ventricular septal defect (VSD) were visible. Autologous glutaraldehyde-treated pericardium was used to reconstruct the aortic-mitral curtain. For repair of the TOF a vertical incision in the right ventricular outflow tract was performed and the infundibular septum was resected. The perimembranous VSD was closed with glutaraldehyde-fixed autologous pericardium, whereas the cranial part of the patch formed the aortic annulus. An aortic homograft was implanted as a full aortic root. The patient recovered early and uneventful. Adult TOF may be complicated by acute aortic valve endocarditis with emergent surgical intervention. Homograft aortic valve replacement is feasible in this setting.

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1. Introduction

Acute endocarditis complicated by aortic valve regurgitation is an uncommon finding in adults with surgical untreated tetralogy of Fallot (TOF). The pulmonary valve is much more likely to be involved in endocarditis in untreated TOF patients than the aortic valve. The hemodynamic consequences for both, the right and left ventricles, are significant. However, the right ventricle may be in a disastrous situation, since a compromised right ventricle from longstanding pressure overload may not tolerate acute volume overload. Thus, emergent surgical intervention is mandatory. Here we report an adult patient with TOF and acute severe aortic valve regurgitation due to bacterial endocarditis with preoperative low cardiac output syndrome (NYHA IV), right atrial pressure of 21 mmHg and dyspnea at rest who was successfully treated with homograft aortic root replacement and correction of TOF.

2. Case presentation

A 28 year male African was admitted to our hospital with congestive right heart failure with clinical symptoms of acute infection and dyspnea at rest (NYHA IV). He had no prior contact to another hospital and had therefore no prior treatment. Transthoracic echocardiography was performed and the diagnosis of TOF was made complicated by moderate aortic valve regurgitation, with floating vegetation attached to the non-coronary aortic valve leaflet. Furthermore, right ventricular (RV) function was severely depressed. Transoesophageal echocardiography (TEE) revealed subaortic abscess cavities. Due to the desperate clinical situation (right atrial pressure 21 mmHg) of the patient he was scheduled for emergent operation.

Median sternotomy was performed and cardiopulmonary bypass was established. The operation was performed under moderate hypothermic cardiopulmonary bypass. For myocardial protection intermittent cold blood cardioplegia was used. After aortotomy of the ascending aorta, a tricuspid aortic valve with large vegetation attached to
the non-coronary aortic leaflet was seen. After excision of the aortic leaflets a large abscess cavity underneath the left and non-coronary sinus and a TOF typical perimembranous ventricular septal defect (VSD) were visible. The abscess cavity was aggressively debrided with resection and curettage of necrotic tissue and povodine swabbing was performed. Autologous glutaraldehyde-treated pericardium was used to reconstruct the aortic-mitral curtain. For repair of the TOF a vertical incision in the right ventricular outflow tract was performed and the infundibular septum was resected. The pulmonary annulus was at lower limit (23 mm). Pulmonary artery incision showed a tricuspid pulmonary valve with macroscopically normal, but rather thin leaflets and normal leaflet coaptation. Due to the thin pulmonary leaflets, we did not consider a Ross procedure with pulmonary autograft and pulmonary homograft in this setting of acute aortic valve endocarditis in a TOF patient. After closing the pulmonary artery the perimembranous VSD was closed with glutaraldehyde-fixed autologous pericardium, whereas the cranial part of the patch formed the aortic annulus. An aortic homograft was implanted as a full aortic root with interrupted sutures to the reconstructed aortic annulus. The coronary arteries were reimplanted and the right ventricular incision was closed with an oval patch of glutaraldehyde treated autologous pericardium. The patient was successfully weaned from cardiopulmonary bypass and simultaneous intraoperative right and left ventricular pressure monitoring showed a $P_{RV/LV}$ of 0.35. In stable hemodynamic condition the patient was extubated on the first postoperative day. Microbiological specimen obtained in the operation revealed a staphylococcus aureus, which was treated according to the antibiogram with a combination of three i.v. antibiotics. After an uneventful postoperative course the patient was discharged from hospital on a 6 weeks course of intravenous antibiotics.

At the latest follow-up the patient presented with NYHA class I and TTE showed normal aortic and pulmonary valve function.

3. Comment

Acute aortic valve endocarditis with subsequent aortic valve regurgitation is a rare clinical finding in patients with untreated TOF. In the literature only few reports have been published dealing with this problem, moreover, the patients reported on were mainly children and less often adolescents [1–5]. The clinical importance of TOF complicated by acute aortic valve regurgitation arises from the hemodynamic changes with acute right ventricular overload on an already compromised RV due to long-standing pressure overload. These changes may result in an acute life threatening situation for the patient. Our patient was initially treated with antibiotics elsewhere, but his clinical situation gradually deteriorated within days and he was subsequently admitted to our hospital with acute right heart failure. We believe that a conservative treatment approach in these patients is not justified even in the presence of moderate aortic valve regurgitation only, since these patients might be prone to acute right heart failure [6,7]. In this situation only immediate surgical intervention may provide an acceptable outcome.

The number of adult patients with untreated TOF has nowadays become a rarity in the western hemisphere, however, in third world countries the number of people with untreated TOF in adolescents and young adults is still high [8,9]. Keeping this in mind and considering the substantial influx of people from third world countries to western countries within the last decade, it is likely that the number of adult patients presenting with untreated TOF and aortic valve endocarditis will increase. Thus, we need a new awareness for these patients who will need earlier surgery in the presence of aortic valve endocarditis complicated by aortic valve regurgitation to prevent acute right heart failure.

References