Thalassemia and Heart Surgery: Aortic Valve Repair After Endocarditis

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Outcome after heart valve surgery in patients affected by thalassemia is an unreported issue and to the best of our knowledge only 7 cases have been described in the literature. Heart valve disease is commonly encountered in thalassemia patients and heart valve replacement carries high risk of prosthesis complications including thrombosis and embolization despite optimal anticoagulation management. We report a successful long-term outcome after a case of aortic valve repair after mycotic valve endocarditis.


The life expectancy of patients affected with thalassemia is significantly prolonged by modern therapy [1], though heart disease still remains the major cause of mortality and morbidity in these patients [1]. Cardiac involvement in thalassemia includes a broad spectrum of diseases. Leaflet thickening, aggressive calcifications, and rupture of the chordae tendineae of the native heart valves reveal congenital elastic tissue disorders, and expose the valve itself to the risk of aggressive infections [2]. Blood transfusions, immune system deficiency, and frequent streptococcal infections can further increase the propensity toward endocarditis and rheumatic valve disease. All these factors can indicate patients for surgical interventions, with several challenges in performing open heart surgery in thalassemia patients [3, 4].

Cardiac valve operations have been anecdotally reported in patients affected with thalassemia [2–7], and with poor long-term follow-up [5, 6] and outcomes [2, 3, 7]. In this article, we add a further case of a thalassemia patient affected with aortic valve endocarditis requiring surgical treatment and compare our outcome with those of all available cardiac valve operations in these patients found in the literature (see Table 1).

A 38-year-old man with β-thalassemia major was referred to our institute for aortic valve endocarditis. A thalassemia genotype IVS1, 110/B 39 was diagnosed 11 months after his birth, and he had received regular blood transfusions every 20 days for the last 11 years, showing a mean hemoglobin level before transfusion ranging from 9 to 9.5 g/dL. Spleenectomy was performed at age of 5 years, and he began iron chelation therapy at the age of 8. He had had a mean ferritin level for the previous 5 years of 300 ng/mL. The last trans-thoracic echocardiogram follow-up, performed 3 years before, showed no cardiac valve abnormalities or disease-related heart involvement. Six months before the diagnosis of endocarditis, the patient experienced a long period of hospitalization because of severe polytrauma complicated by a persistent fever and Candida parapsilosis bloodstream infection. Due to the persistency of the infection in the blood despite voriconazole therapy, a transesophageal echocardiogram was performed and showed 3 floating vegetations, ranging from 5 to 15 mm, involving the aortic side of the aortic valve leaflets, and moderate aortic regurgitation (Fig 1A). Antifungal treatment was switched to liposomal amphotericin B combined with caspofungin for a total of 8 weeks after the diagnosis. Given the fungal infection, high embolic risk, and the persistent septic status the patient was advised to undergo aortic valve surgery.

Preoperative investigation revealed hemoglobin and hematocrit levels of 9 g/dL and 29.7%, respectively, and computed tomographic findings were unremarkable. A standard surgical and anesthetic procedure was done. On examination a preserved trileaflet aortic valve was detected, and the vegetations gently removed (Fig 1B). The loss of coaptation between the noncoronary and the right coronary

Table 1. Review of Thalassemia and Heart Surgery in the Literature

<table>
<thead>
<tr>
<th>Author</th>
<th>Disease</th>
<th>Surgery</th>
<th>Complications and Follow-Up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Omoto</td>
<td>AR</td>
<td>AVR, mechanical</td>
<td>6 months</td>
</tr>
<tr>
<td>Darwazah [2]</td>
<td>MVE, Gram-</td>
<td>MVR, mechanical</td>
<td>Early PO death</td>
</tr>
<tr>
<td>Botta</td>
<td>AS</td>
<td>MVR, mechanical</td>
<td>6 months</td>
</tr>
<tr>
<td>Farmakis [7]</td>
<td>AS</td>
<td>AVR, mechanical</td>
<td>Prosthetic valve thrombosis; 16 months</td>
</tr>
<tr>
<td>Metras</td>
<td>MR</td>
<td>MVRep</td>
<td>–</td>
</tr>
<tr>
<td>deLeval [3]#</td>
<td>–</td>
<td>MVRs, biologic</td>
<td>Recurrent systemic embol, reoperation; 1 to 4 years</td>
</tr>
</tbody>
</table>

# Requiring mechanical AVR reoperation and complicated by disseminated coagulopathy and death.  * Two patients.

AR = aortic regurgitation; AS = aortic stenosis; AVR = aortic valve replacement; MR = mitral regurgitation; MVE = mitral valve endocarditis; MVR = mitral valve replacement; MVRep = mitral valve repair; MVRs = mitral valve replacement(s); PO = postoperative.

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cups due to annular dilatation was corrected by subannular commissuroplasty using Ethibond 2/0 (Ethicon, Somerville, NJ) pledged with autologous pericardium (Fig 1C). In order to reduce the risk of hemolysis and red blood cell trauma we performed a normothermic cardiopulmonary bypass, antegrade blood cardioplegia, and decreased suction power. The pledget was obtained by autologous pericardium. Transfusions of 3 units of packed red blood cells became necessary. The recovery was completely uneventful and the patient was discharged home after 14 days. Anticoagulation was achieved with a month of subcutaneous heparin, with no antiplatelet drugs used in the long-term period. Three years after the operation the patient is in good clinical condition, with no relapse of Candida parapsilosis infection. A transthoracic echocardiogram showed a good outcome of the aortic valve repair technique (Fig 1D).

Comment

Cooley’s disease or thalassemia major is the most severe form of thalassemia (derived from the Greek: “thalassa,” meaning “the sea” and “emia,” meaning “related to the blood”) with a β-gene of hemoglobin anomaly of greater than 1% in the Mediterranean Basin [1]. The quality of life for these patients is extremely poor due to the need for continual transfusions, and hospital admission due mainly to heart and infectious disease [1–8]. Given the prolonged survival and the heart involvement of the disease, cardiac operations can be required though there are few reports in the literature of cardiac surgery in these patients.

First described in 1974 [3], cardiac valve surgery in thalassemia patients poses important issues because of cardiopulmonary complications and management [8]. The use of extracorporeal circulation can result in increased hemolysis of fragile erythrocytes [3, 4], and appropriate measures should be advocated [8].

Our case adds to the literature concerning patients affected with thalassemia and requiring cardiac valve surgery. The repair technique offered a substantial advantage to the patient in long-term survival, freedom from cardiac events, and recurrent infection. Despite reports of success [5–7], valve replacement with a mechanical prosthesis should be considered with caution because of the worsening of chronic intravascular hemolysis and thrombotic propensity [7]. The hypercoagulable status is due to platelet and endothelium activation, membrane changes in red blood cells, and low levels of antithrombin III, protein C, and protein S [7]. On the other hand, the biologic prosthesis carries an increased risk of thromboembolism and reoperation [3] also because of the young age of these patients requiring heart valve surgery.

This is the first report of mycotic infection involving the aortic valve in β-thalassemia patients. The endocarditis affected a structurally normal aortic valve, suggesting an aggressive course of fungal pathogens in severely immunocompromised and prolonged hospital stay patients with long-term indwelling central venous catheter. Candida parapsilosis is associated with nosocomial infections and invasive candidiasis, and the native aortic valve is the most commonly involved (42.5%).

In conclusion, conservative valve surgery has to be considered the first surgical choice in patients affected with β-thalassemia, even if the native valve function can be impaired by the elastic tissue disruption caused by the disease [7]. Compared with the few reports in the literature, our patient had a satisfactory long-term outcome, including freedom from recurrent valve endocarditis.

References