

Infectious endocarditis of the mitral valve with hypertrophic cardiomyopathy in Costello syndrome

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Abstract

Costello syndrome is a rare congenital disease caused by activating germline mutations, and it is often associated with cardiac abnormalities. A 17-year-old male, who had a history of Costello syndrome, presented with persistent fever. The vegetation attached to anterior mitral leaflet was detected, and antibiotic therapy was administered as treatment for infectious endocarditis. However, it was difficult to manage his heart failure owing to the worsening of mitral valve regurgitation. Therefore, mitral valve repair with vegetation resection was performed. His hypertrophic cardiomyopathy and systolic anterior motion caused left ventricle outflow tract obstruction. A floating stitch on the anterior mitral leaflet from the posterior ring annulus was effective. Herein, we report a successfully repaired case of infectious endocarditis on the mitral valve with hypertrophic cardiomyopathy complicated by Costello syndrome.

Title: Infectious endocarditis of the mitral valve with hypertrophic cardiomyopathy in Costello syndrome

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Short running title: Infectious endocarditis in Costello syndrome

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Abstract:

Costello syndrome is a rare congenital disease caused by activating germline mutations, and it is often associated with cardiac abnormalities. A 17-year-old male, who had a history of Costello syndrome, presented with persistent fever. The vegetation attached to anterior mitral leaflet was detected, and antibiotic therapy was administered as treatment for infectious endocarditis. However, it was difficult to manage his heart failure owing to the worsening of mitral valve regurgitation. Therefore, mitral valve repair with vegetation resection was performed. His hypertrophic cardiomyopathy and systolic anterior motion caused left ventricle outflow tract obstruction. A floating stitch on the anterior mitral leaflet from the posterior ring annulus was

effective. Herein, we report a successfully repaired case of infectious endocarditis on the mitral valve with hypertrophic cardiomyopathy complicated by Costello syndrome.

Introduction:

Costello syndrome (CS) is a rare congenital disease caused by activating germline mutations.¹ It is characterized by deficient postnatal development of multiple organs, mental retardation, and predisposition to cancer. Congenital malformations such as pulmonary valve stenosis, hypertrophic cardiomyopathy (HCM), and atrial septal defect have been reported.² Infectious endocarditis in a patient with CS has not been previously reported. We herein present the rare case of a 17-year-old male with CS who developed mitral valve regurgitation (MR) because of infectious endocarditis.

Case Report:

A 17-year-old male presented with fever of unknown origin and was admitted for examination and treatment. He had a history of CS and atopic dermatitis as well as mental retardation according to CS. Blood cultures detected methicillin-susceptible *Staphylococcus aureus*. Echocardiography showed HCM and vegetation attached to the anterior mitral leaflet (AML) with MR. MR was less than mild and severe symptoms of heart failure were not present. Therefore, antibiotic and medication therapy for heart failure were initiated. Although the infection responded to treatment, the MR and heart failure symptoms worsened. Consequently, surgery for vegetation resection and mitral valve repair was planned.

After median sternotomy, cardiopulmonary bypass was established with ascending aortic cannulation and bicaval drainage, and the heart was arrested with antegrade cardioplegia. The mitral valve was visualized through a right-sided left atriotomy. After shaving the vegetation on the AML, mitral valve annuloplasty was performed using a 25-mm Tailor Flexible Annuloplasty Ring (St. Jude Medical, St. Paul, MN, USA) with single-pair artificial chordae reconstruction using CV4 Gore-Tex suture (WL Gore and Associates, Inc., Flagstaff, AZ, USA). Transesophageal echocardiography showed systolic anterior motion (SAM) of the mitral valve with moderate MR and left ventricular outflow tract obstruction (LVOTO) owing to HCM after unclamping the aorta (Figure 1). To reduce SAM, the floating stitch technique was used:³ a double-arm CV4 Gore-Tex suture was applied just to the middle of the tip (A2) of the AML in a figure-eight fashion, and both arms of the suture were fixed to the annuloplasty ring at the middle of the posterior annulus (P2) (Figure 2). After application and fixation, SAM and MR disappeared (Figure 3). The patient's postoperative course was uneventful, and he was later discharged home.

Discussion:

CS is often associated with multiple growth retardation and malformation in multiple organs, including cardiac abnormalities. In this case, remarkable complication interfering with a surgery was HCM, and the ascending aorta was small relative to his physique. In addition, the disorder of connective tissue and deposition of elastic fiber was one of the characteristics of CS, and the elastic fibers of aorta and coronary arteries are thinner in patients with CS than in normal individuals.⁴ Therefore, detailed preoperative examination and careful technique are required in these patients. We used a flexible ring because of the irregularly shaped annulus. To manage LVOTO, the floating stitch technique was used to reduce SAM of the mitral valve after valve repair.³ This method is a simple, reproducible, and effective for preventing SAM. Further follow-up and studies on this technique are needed.

Conclusion:

In patients with CS with associated cardiac and tissue abnormalities, mitral valve repair using a flexible ring and artificial chordae reconstruction using the floating stitch technique to manage LVOTO are feasible.

Conflicts of Interest: The authors declare that there are no conflict of interests.

Author contributions:

S.I., M.M., T.F. and K.Y. designed and performed the experiments, analyzed data and interpreted it. S.I.

and K.Y. Drafted article. S.I., M.M., F.T. and K.Y. revised it critically. S.I., M.M., F.T. and K.Y. approved of the article, collected data and supported technical and logistical.

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Figure.1 Systolic anterior motion of anterior mitral leaflet (arrow) developed and it caused mitral valve regurgitation after the initial mitral valve repair. LA, left atrium; LV, left ventricle; AO, aorta

Figure.2 Intraoperative images from the surgeon's perspective showing the floating stitch technique. A double-arm CV4 Gore-Tex suture (arrows) is applied just to the middle of the tip (A2) of anterior mitral leaflet (AML) in a figure-eight fashion, and both arms of the suture are fixed to the annuloplasty ring (asterisk) at the middle of the posterior annulus (P2).

Figure.3 Postoperative transthoracic echocardiography. A, The anterior leaflet (red arrow) was retracted to the posterior annulus by Gore-tex suture during diastole. Mitral stenosis did not exist. B, Systolic anterior motion was not shown, and the anterior leaflet was in a closed position (yellow arrow) during systole. LA, left atrium; LV, left ventricle; AO, aorta



