Pulmonary Valve Bacterial Endocarditis in Tetralogy of Fallot

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Received 27 August 2006; Accepted 22 October 2006

Abstract

We report two cases of Tetralogy of Fallot with pulmonary valve bacterial endocarditis where one extended to the branch of pulmonary artery (PA). This is a rare occurrence. Aggressive supportive care plus early and radical surgery can be life saving.

Keywords: Tetralogy of Fallot • Pulmonary valve endocarditis • Congenital heart disease

Introduction

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease.1-3 Infective endocarditis is uncommon condition with a high probability of morbidity and mortality in association with congenital cardiac malformations.4 Bacterial endocarditis is a serious and fatal complication in congenital heart disease5,6 and is a rare complication of TOF. In one report,7 TOF was seen only in 9.5% of cases undergoing surgical treatment for infective endocarditis associated with congenital heart disease. In these cases, endocarditis involved tricuspid valve and ventricular septal defect (VSD), but isolated pulmonary valve bacterial endocarditis has not been reported.

Case Reports

Case 1

A 7-year-old girl presented with a one month history of fever, dyspnea, aggravated cyanosis and weight loss. She was a known case of TOF since birth but was never treated before. She received antibiotic therapy one week prior to admission. Physical examination revealed blood pressure, 90/50 mmHg; heart rate, 120/min and temperature, 38.3ºC. Other findings were systemic cyanosis, pulmonary crackle and clubbing. Laboratory examination showed hemoglobin concentration (Hb) 17.9 g/dl, white blood cell count (WBC) 16.800/mm³ and sedimentation rate 84mm/hour. Chest X-Ray depicted an enlarged heart and two pulmonary consolidations in the right lower pulmonary lobe.

Echocardiography revealed large VSD, overriding of aorta, normal left ventricular ejection fraction (LVEF), right ventricular outflow tract (RVOT) obstruction with 78mmHg gradient, small pulmonary annulus, and no visible pulmonary valve with large vegetation (3.5× 1cm). This vegetation extended to the bifurcation of pulmonary artery (figure 1). Multiple blood culture was negative. Brain CT scan was normal, and abdominal CT scan showed splenomegaly.

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After 72 hours of aggressive empiric antibiotic therapy, emergency surgery was done (due to fear of massive pulmonary emboli). At surgery, large vegetation filled main pulmonary trunk was found. After complete evacuation of vegetation and debridment of necrotic tissue including pulmonary annulus, VSD was closed by a Gore-Tex patch. Subvalvular resection was done and main pulmonary artery replaced by number 21 pulmonary homograft. Right and left branches and bifurcation of PA was enlarged by glutaraldehyde treated pericardium (figure 2). The postoperative course was uneventful. Echocardiography revealed 10mmHg gradient across homograft with no residual VSD. Vegetation culture was positive for Staphylococcus Aureus. The patient was treated with appropriate antibiotics for an additional four weeks.

Case 2

A 19-year-old girl presented with fever and dyspnea for 5 weeks. She was a known case of TOF but neglected as a remote rural area inhabitant.

She was treated for lung abscess in another center and referred after 4 a week course of antibiotic therapy and percutaneous drainage of lung abscess. She had extreme respiratory distress when admitted to our center, so she was intubated and respiratory support was launched. Diagnostic workup began immediately.

Physical examination revealed dehydration, blood pressure, 100/70mmHg; pulse rate, 130/min; temperature, 38°C; central cyanosis, clubbing, respiratory crackle and rhonchi, ascitis and leg edema. Laboratory examination showed: HB, 11.3 g/dl; WBC, 8100/mm³; Creatinine, 2.0 mg/dl and blood urea nitrogen, 94 mg/dl. Chest X-Ray revealed multiple pulmonary infiltrations especially in sub plural region of both lungs.

Echocardiography revealed large perimembrane VSD, overriding of aorta, severe valvar and sub valvular pulmonary stenosis, multiple vegetations on pulmonary valve and RVOT, 2-3 plus tricuspid valve (TV) regurgitation and TV annular dilatation (figure 3).

After two days of stabilization and antibiotic therapy, the patient was operated. During operation, multiple large vegetations were observed on RVOT, pulmonary valve, main PA and branches of PA. The PA bifurcation, main PA, pulmonary annulus and endocardium of RVOT was resected. PA was reconstructed by a bifurcated pulmonary homograft (No. 23) and pericardial patch, VSD closure was performed by Gore-Tex patch. Tricuspid valvoplasty was done (figure 4).
Infective endocarditis was proven by clinical status and pathological findings. The postoperative care was complicated with sepsis and respiratory failure. After 6 days of supportive therapy, her respiratory and general condition improved and the patient was extubated at day 7. Intravenous antibiotic therapy continued for an additional 6 weeks. Oral antibiotic followed for an additional 2 months. Follow-up echocardiography showed perfect repair and no evidence of intracardiac infection. All cultures were negative. In chest X-Ray, pulmonary lesions had healed.

Conclusion

Isolated pulmonary valve bacterial endocarditis complicating Tetralogy of Fallot is very rare. This complication is fatal. There is minimal guideline for accurate treatment. In our experience appropriate supportive care, early aggressive surgery (debridment of all necrotic and inflamed tissue) and use of homograft is applicable with good results.

References