CASE REPORT

Ross Procedure for Congenital Aortic Insufficiency and an Associated Right Intramural Coronary Artery

A 7-year-old boy diagnosed with aortic insufficiency was treated with a Ross procedure and he had an associated right intramural coronary artery. Although preoperative angiocardiography showed that the right and left coronary artery orifices were close together, a right intramural coronary artery could not be diagnosed by transthoracic echocardiography. Intraoperative findings showed that the right coronary artery was intramural for a distance of 7 mm. As a result, a longer single coronary button was harvested and transplanted to the pulmonary autograft. The postoperative course was uneventful; coronary ischemia did not occur and aortic valve function was preserved.

Key words: Ross procedure, intramural coronary artery, single coronary button, aortic insufficiency

Shin Takabayashi, MD, Hideto Shimpo, MD, and Yoichiro Miyake, MD.

Anomalies of the coronary arteries (CA) occur very rarely. Congenital heart defects, such as transposition of the great arteries (TGA), tetralogy of Fallot, pulmonary atresia may accompany this rare lesion. The most common cause of acquired abnormalities of the CA is Kawasaki disease. Congenital aortic valvular diseases with an associated intramural coronary artery (IMCA) have not been reported previously. In this case report, we describe a patient who underwent a successful Ross procedure with a longer single coronary button transplantation for congenital aortic insufficiency (AI) with a right IMCA.

Case

A 7-year-old boy was referred to us for surgical evaluation. His height was 119 cm and weight was 20.5 kg. At 1 month of age, he was diagnosed with congenital aortic stenosis (AS), and underwent percutaneous balloon aortic valvuloplasty. Because of residual stenosis, surgical aortic valve commissurotomy was performed at 7 months of age. His AI had progressed, prompting his referral to us. Echocardiography confirmed the presence of severe AI and mild AS. The aortic valve was thickened, and the aortic valve annulus was 16 mm and pulmonary valve annulus was 17 mm in diameter. The aortic valve was bicuspid without subvalvular stenosis. An IMCA was not detected by transthoracic echocardiography. Electrocardiography (ECG) showed a strain pattern in V5 and V6, indicating progressive left ventricular volume overload. At the time of catheterization, the pressure gradient between the left ventricle (LV) and the ascending aorta was 38 mmHg and the left ventricular endodiastolic pressure was 15 mmHg. Angiography revealed severe AI and poststenotic dilation of the aorta. Although aortography showed that the right and left coronary orifices were very close together, a right IMCA could not be diagnosed (Fig. 1). A history of unexplained syncope or chest pain suggesting a congenital coronary anomaly was not elicited preoperatively.

The operative findings revealed that the aortic valve was bicuspid and thick. The right and left CA originated very close together although there was not a single CA. The right CA was intramural for a distance of 7 mm from the orifice to anterior to the aorta (Fig. 2A). Both the right and left coronary orifices were harvested as a single coronary button that was made longer anteriorly.
along the right IMCA, and transplanted to the pulmonary autograft (Fig. 2B). Right ventricular outflow tract reconstruction was performed using a hand-fashioned ePTFE valved conduit 24 mm in diameter.

Postoperatively, the patient was extubated the day after surgery and the duration of the intensive care unit stay was 2 days. Coronary ischemia did not occur, and the hospital stay was 26 days. Follow-up echocardiography showed no neoaortic insufficiency and no neoaortic cusp distortion or left ventricular outflow tract stenosis. Follow-up ECG showed no evidence of coronary ischemia or cardiac infarction. The patient is alive and well without coronary ischemia or progressive aortic lesions after a 21 month follow-up period.

Discussion

Congenital anomalies of the CA are rare and can cause syncope, myocardial infarction, and sudden death. In patients with TGA, detailed studies of the anatomy of the CA have been performed and demonstrate that the frequency of an IMCA is 0.8%. Furthermore, patients with an IMCA had a greater mortality. Use of the Ross procedure for congenital aortic lesions with an associ-