Three-vessel Coronary Artery Disease Complicated with Congestive Heart Failure in a Highly Aged Patient with Tetralogy of Fallot Having Undergone Palliative Surgeries

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Abstract

An increasing number of patients with tetralogy of Fallot (TOF) are reaching older age. We encountered a 75-year-old woman with uncorrected TOF and concomitant severe coronary artery disease (CAD) with congestive heart failure. Her CAD risk factor was hyperlipidemia, which had been untreated. Successful percutaneous coronary interventions have improved her clinical condition and provided long-term survival. Although CAD is considered to be a rare complication in adults with TOF, both strict modification of CAD risk factors and early detection of CAD would be also required in this population, given the residual TOF lesions relating to acute exacerbation of clinical presentation.

Key words: congenital heart disease, congestive heart failure, coronary artery disease, percutaneous coronary intervention, rotational atherectomy, tetralogy of Fallot

Introduction

An increasing number of patients with tetralogy of Fallot (TOF) are reaching older age, since the first successful intracardiac repair of TOF in 1954 (1). It is suggested that adults with cyanotic congenital heart disease, including TOF, lack coronary atherosclerosis and have low cholesterol levels (2, 3), although the incidence of coronary artery disease (CAD) has not been established in this patient population. In contrast, only a few cases with concomitant CAD in the elderly survivors with TOF have been reported (4-7). Here, we describe a 75-year-old woman with TOF having undergone palliative surgeries, who developed into severe three-vessel CAD complicated with congestive heart failure. In addition, we discuss clinical management in this patient population.

Case Report

A 75-year-old woman with TOF, who had undergone palliative operations, Blalock-Taussig shunt surgeries, at age 35 and 52 separately, was admitted to our department with orthopnea in June 2003. Chest x-ray showed significant pulmonary edema with massive bilateral pleural effusion (Fig. 1A). Twelve-lead electrocardiogram (ECG) revealed poor R-wave progression in the precordial leads and mild ST segment depression in the limb and left precordial leads, all of which were similar findings to the previous ECG recordings (Fig. 1C). The examination of blood biochemistry including cardiac enzymes on admission revealed no abnormal findings, except for the elevation of serum brain natriuretic peptide (202 pg/ml). Given those findings in ECG and blood biochemistry, it was not thought that she had an acute or impending myocardial infarction. Transthoracic echocardiogram showed a 30 mm ventricular septal defect with bidirectional shunting, an overriding aorta, subvalvular pulmonary stenosis, right ventricular hypertrophy with normal right ventricular wall motion, and left ventricular anteroseptal hypokinesis with an ejection fraction of 52% (Figs. 1D, 1E).

Two weeks later, the usual medical therapy for congestive heart failure improved her clinical status, as pulmonary
edema disappeared (Fig. 1B). Still after the amelioration of congestive heart failure, the examination of arterial blood gas inherently revealed chronic hypoxemia due to her uncorrected TOF, as expected: the partial pressure of arterial oxygen was 51 mmHg, the partial pressure of arterial carbon dioxide was 44 mmHg, and the oxygen saturation was 87% in a room-air condition, respectively. She usually appeared to be mildly cyanotic, however, she had no erythrocytosis. The results of both the hematologic test including hematocrit and platelet counts and the coagulation study were all normal. It was plausible that the occurrence of congestive heart failure might be attributed, at least in part, to the occlusions of the Blalock-Taussig shunts. However, chest computed tomography demonstrated that the Blalock-Taussig shunt on the right side retained the vascular patency, although that on the left side was totally occluded (Fig. 2). Thereafter, during the hospital stay, she had shortness of breath on exertion in accordance with ST segment depression in the left precordial
Figure 2. A, Three-dimensional computed tomography showing pulmonary arteries and veins, and great arteries in the view from her back. Arrow indicates the right Blalock-Taussig shunt, and arrowhead indicates brachiocephalic artery. Ao = aorta. B and C, Two-dimensional contrast computed tomography of the chest revealed that the Blalock-Taussig shunt on the left side was totally occluded (arrowhead), but that on the right side retained the vascular patency (arrows).

Figure 3. A, Right ventriculogram revealed infundibular subvalvular pulmonary stenosis (arrowhead) and the concomitant left ventriculogram via ventricular septal defect, suggesting the existence of right-to-left shunt. B, C, and D, Coronary angiogram showed severe stenotic lesions with calcification in the distal right coronary artery (RCA) (B; arrow), in the mid-portions of the left circumflex coronary artery (LCX) (C; arrow and arrowhead), and in the mid-portion of the left anterior descending artery (LAD) (D; arrow). In contrast, coronary ectasia, which is considered to be characteristic of patients with cyanotic congenital heart disease, was hardly detected. E and F, Intravascular ultrasound imaging before intervention showed that both the LCX (C; arrow) and LAD (D; arrow) lesions were exhibiting the superficial calcified plaques and vessel shrinkage. G, H, I, J, and K, Coronary angiogram (G, H, I) and intravascular ultrasound imaging (J, K) after the successful percutaneous coronary interventions showed the optimal dilatations in all lesion sites of the three coronary arteries. The percutaneous coronary interventions were composed of both stenting following balloon angioplasty in the RCA and LCX lesions and stenting following the debulking by rotational atherectomy (burr size: 1.75 mm) in the diffuse LAD lesion.
leads, suggesting the concomitant ischemic heart disease. Therefore, she then underwent cardiac catheterization.

Right heart catheterization demonstrated the equivalent between right and left ventricular pressure patterns, as both ventricular systolic pressures were 130 mm Hg. The step-up of oxygen saturation was detected from right atrium to right ventricle (55% to 61%), suggesting the existence of left-to-right shunt. Unfortunately, the catheters could not be advanced into pulmonary artery due to severe subvalvular pulmonary stenosis. Consequently, pulmonary arterial pressure was undefined, and the oximetry study turned out to be incomplete such as the pulmonary to systemic flow ratio (Qp/Qs) and the right-to-left shunt rate could not be precisely calculated. Next, right ventriculogram demonstrated infundibular subvalvular pulmonary stenosis and the concomitant left ventriculogram via ventricular septal defect, suggesting the existence of right-to-left shunt (Fig. 3A). Coronary angiogram subsequently showed severe stenotic lesions with calcification in the distal right coronary artery (RCA), in the mid-portions of the left circumflex coronary artery (LCX), and in the mid-portion of the left anterior descending artery (LAD), whereas coronary ectasia, which is considered to be characteristic of patients with cyanotic congenital heart disease (2, 3), was hardly detected (Figs. 3B, 3C, 3D). Accordingly, the major etiology of congestive heart failure in this patient was considered as progressive three-vessel CAD. Except for aging, her CAD risk factor was only hyperlipidemia (serum total cholesterol of 262 mg/dl), which had been untreated in the out-patient clinic. Fortunately, this patient has obtained good long-term clinical outcome owing to successful percutaneous coronary interventions for the severe three-vessel CAD.

The recent studies have reported that hypoxemic erythrocytotic adults with cyanotic congenital heart disease, including TOF, lack coronary atherosclerosis, mimicking hypoxemic erythrocytotic residents of high altitudes (2, 3). It has been suggested that the low incidence of coronary atherosclerosis in them might be attributed to hypocholesterolemia, hypoxemia, upregulated nitric oxide, hyperbilirubinemia, and low platelet counts, many of which are mainly derived from secondary erythrocytosis. Those studies (2, 3) revealed that inherently cyanotic patients with congenital heart disease had significantly lower total cholesterol levels even after the surgical elimination of cyanosis, whereas inherently aycnotic patients with congenital heart disease had the usual total cholesterol levels similar to those predicted by the Framingham study (10). However, it was concurrently revealed in those papers (2, 3) that a few patients with cyanotic congenital heart disease had significantly elevated cholesterol levels, such as the present case, and that there were no apparent differences in family history or dietary habits between these few cyanotic patients with elevated cholesterol levels and most cyanotic patients with low cholesterol levels. Those findings have suggested that cyanotic patients with congenital heart disease consist of a heterogeneous population, even concerning lipid profile, and that the examinations of blood biochemistry concerning lipid profile are also mandatory in this patient population. The present patient with uncorrected TOF certainly had mild cyanosis and hypoxemia, which are considered to be antiatherogenic factors, but she had no erythrocytosis. Moreover, neither hyperbilirubinemia nor low platelet counts were detected. Given those findings in addition to her hypercholesterolemia, it may be no wonder that she developed into severe CAD with age.

Coutu and colleagues have reported that TOF patients with concomitant CAD are characterized by urgent clinical presentation, worsening hemodynamics, and poor control of CAD risk factors (7), which are exactly applicable to the present case. Urgent clinical presentation may be associated with the residual TOF lesions (i.e., ventricular septal defect). In the present case with severe three-vessel CAD, the main pathological condition was acute congestive heart failure without the preceding typical anginal pain. The residual ventricular septal defect with right-to-left shunt in this case might augment significantly the adverse effect of myocardial ischemia, leading to an acute onset of congestive heart failure. Given the residual TOF lesions deeply relating to acute exacerbation of clinical presentation, both strict modification of CAD risk factors and early detection of CAD would be also required in adults with TOF, particularly, in the elderly population, even concerning lipid profile, and that the examinations of blood biochemistry concerning lipid profile are also mandatory in this patient population. The present patient with uncorrected TOF certainly had mild cyanosis and hypoxemia, which are considered to be antiatherogenic factors, but she had no erythrocytosis. Moreover, neither hyperbilirubinemia nor low platelet counts were detected. Given those findings in addition to her hypercholesterolemia, it may be no wonder that she developed into severe CAD with age.

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Thus far, there have seldom been patients with corrected or uncorrected TOF who reach the seventh decade (8, 9). In addition, CAD has been considered as a rare complication in adults with TOF, and indeed, the present case is one of the oldest patients with TOF and concomitant CAD in the literature (4-7). Before the occurrence of congestive heart failure, her CAD risk factor, hyperlipidemia, which might contribute to the progression of the severe three-vessel CAD with age, had been untreated in the out-patient clinic. Fortunately, this patient has obtained good long-term clinical outcome owing to successful percutaneous coronary interventions for the severe three-vessel CAD.

Discussion

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survivors with uncorrected TOF, even if CAD might be a relatively rare complication in this patient population.

In conclusion, we encountered a 75-year-old woman with uncorrected TOF and concomitant severe CAD along with acute congestive heart failure. A careful observation and treatment for both CAD and its risk factors must be continued in this case hereafter. Although CAD is usually considered to be a rare complication in adults with TOF, we might need to keep in mind the possibility of the concurrent occurrence of CAD in the elderly patients with TOF, who have recently been increasing.

References