Fontan operation in a paediatric patient with a history of Takotsubo cardiomyopathy

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Received 19 December 2013; received in revised form 22 February 2014; accepted 4 March 2014

Abstract

Takotsubo cardiomyopathy is very rare in the paediatric population and has not been described in a single-ventricle patient yet. We report the case of a 4-year old boy with a history of Takotsubo cardiomyopathy in whom we performed a Fontan operation. After coil embolization of the minor aortopulmonary collateral arteries, the patient developed Takotsubo cardiomyopathy. His cardiac function largely recovered over 3 months. He subsequently progressed to a Fontan operation and was weaned uneventfully off cardiopulmonary bypass, on minimal doses of dopamine and milrinone; he was sedated using a dexmedetomidine infusion and a midazolam bolus. There were no signs of recurrent Takotsubo cardiomyopathy over the subsequent 2 years.

Keywords: Takotsubo cardiomyopathy • Paediatric patient • Fontan candidate

INTRODUCTION

Takotsubo cardiomyopathy is characterized by transient, severe systolic dysfunction of the apical ventricular segments and typically occurs in women >60 years old, but is very rare in the paediatric population. To our knowledge, single-ventricle disease has not been previously reported. Thus, the safety of the cardiac surgery and risks of recurrence after the surgery are unclear. We report the case of a 4-year old boy with a history of Takotsubo cardiomyopathy in whom we performed the Fontan operation.

CASE

A 4-year old boy was diagnosed with dextrocardia, a single right ventricle, common atrioventricular valve and pulmonary atresia. He was performed a left modified Blalock-Taussig shunt at 1 month of age, a right modified Blalock-Taussig shunt at 2 years of age and a bidirectional Glenn operation and pulmonary artery plasty at 3 years of age. He was admitted to our hospital for coil embolization of the minor aortopulmonary collateral arteries and Fontan operation. The day after the coil embolization, he complained of chest pain, and a physical examination revealed tachypnoea, tachycardia and a gallop rhythm. His electrocardiogram showed ST-segment elevation in the V1–V6 leads and a T-wave inversion in lead III (Fig. 1). The patient’s cardiac enzymes were within the normal ranges, and his serum was negative for troponin T; however, his brain natriuretic peptide (BNP) level was elevated (844.8 pg/ml). An echocardiogram and a ventriculogram revealed akinesis motion of the ventricular apex (Fig. 2A and B) and an estimated ejection fraction of 31%. Further, coronary angiography showed no stenotic or occlusive lesions. From these findings, we diagnosed Takotsubo cardiomyopathy. He was treated for acute heart failure with dobutamine, and 2 days later, his echocardiographic wall motion improved and his BNP level decreased. Three months later, the cardiac function almost completely normalized, his echocardiogram revealed an estimated ejection fraction of 54%, his electrocardiogram showed no signs of ST-segment elevation and T-wave inversion, his laboratory values were within normal ranges and, therefore, Fontan operation was planned.

The extracardiac conduit Fontan operation was performed under general anaesthesia using remifentanil and sevoflurane, with the depth of anaesthesia controlled by bispectral index monitoring; his cardiac function was monitored using a transoesophageal echocardiogram. A median resternotomy was performed and a cardiopulmonary bypass (CPB) was established. The operation was performed on the beating heart. The weaning from CPB was uneventful; 3 µg/kg/min of dopamine and 0.3 µg/kg/min of milrinone were used for haemodynamic support and modified ultrafiltration was performed. Postoperatively, he was sedated with an infusion of dexmedetomidine (0.4 µg/kg/h) and a bolus of midazolam. The patient was weaned from inotropic support after 3 postoperative days. The postoperative course was uneventful, except that he developed a chylothorax. Echocardiograms indicated good ventricular contractions, with an estimated ejection fraction of 57.5%. In the 2 years, since the Fontan operation, there have been no signs of recurrent Takotsubo cardiomyopathy.

DISCUSSION

Takotsubo cardiomyopathy was first described by Satoh et al. in 1990. The disease is characterized by transient, severe systolic dysfunction of the apical ventricular segments and is triggered by...
physical or emotional stress. This cardiomyopathy typically occurs in individuals >60 years old [1], with only 13 paediatric cases found in a PubMed search. To our knowledge, single-ventricle disease has not been previously reported. Typically, over 80% of patients present with chest pain, and their echocardiograms show ST-segment elevations and T-wave inversions, mimicking acute coronary syndrome. However, the cardiac enzymes are typically within the normal ranges or only slightly elevated; coronary angiography reveals the absence of stenotic or occlusive lesions [2]. The pathophysiology of the disease remains unknown; however, it has been suggested that endogenous catecholamines play an important role. Lyon et al. [3] suggested that high concentrations of circulating epinephrine change beta-adrenoceptor coupling from Gs-protein to Gi-protein. The beta-adrenoceptors are usually coupled with the Gs-protein, which produces positive inotropic effects by activating adenylyl cyclases and increasing intracellular cyclic adenosine monophosphate concentrations. In contrast, the Gi-protein produces a negative inotropic effect that suppresses adenylyl cyclases. As beta-adrenoceptors are more often found in apical cardiomyocytes than in basal cardiomyocytes, the apical segments show specific, decreased contraction in Takotsubo cardiomyopathy. Other pathophysiological views suggest, for example, that high concentrations of norepinephrine are released from the sympathetic nerves and lead to microvascular spasms in the coronary circulation that induce apical cardiomyocyte stunning.

In Takotsubo cardiomyopathy, cardiac function generally normalizes within several days to several weeks. Some patients developed serious heart failure and/or arrhythmia, and require...
haemodynamic support such as intravenous catecholamine infusion and mechanical support, including intra-arterial balloon pumping and/or cardiopulmonary support systems. The in-hospital mortality rate has been suggested to be 1.1–1.7% [4], with a recurrence rate of 4–11% [2]. Only one report has described the cardiac surgery in a patient with a history of Takotsubo cardiomyopathy [5]. Thus, the safety and risk of recurrence after cardiac surgery remain largely unclear. This led to a concern that the Fontan procedure might fail owing to recurrence of Takotsubo cardiomyopathy. In this case, the Fontan operation was safely performed on the heart, with appropriate sedation, using low concentrations of catecholamines, and no recurrence has been observed during 2 years of follow-up.

CONCLUSION

We performed the Fontan operation on a patient with a history of Takotsubo cardiomyopathy. Because Takotsubo cardiomyopathy may be triggered by emotional or physical stress, patients need to be carefully sedated and receive appropriate circulatory support to avoid recurrence. With the increasing number of reports of Takotsubo cardiomyopathy, additional cases of cardiac surgery in patients with a history of Takotsubo cardiomyopathy need to be reported to better understand its perioperative management.

Conflict of interest: none declared.

REFERENCES