Myocardial infarction due to thrombosis of native aorta late after Fontan procedure for hypoplastic left heart syndrome

Bhavi Patel, Tacy Downing, Lourdes Prieto, Darline Santana-Acosta
Pediatric Cardiac Critical Care/Cardiology, Nicklaus Children's Hospital, Miami, FL, USA

ABSTRACT

Patients with single ventricle physiology who have undergone the Fontan procedure are at risk for long-term complications such as thrombus formation. Thrombus formation in the native aortic root (NAR) can be life-threatening if retrograde filling of the coronary arteries (CAs) is impaired. We present three cases of NAR thrombus with embolization to the CA, resulting in myocardial infarction. Thromboembolic events are well-documented complications of Fontan physiology, and optimal prevention and treatment strategies are poorly defined.

Keywords: Fontan, hypoplastic left heart syndrome, myocardial infarction, native aortic root thrombus, thromboembolism

INTRODUCTION

We present a series of three adolescent patients with hypoplastic left heart syndrome (HLHS) and Fontan operation, presenting within a 2-year period with acute clinical symptoms consistent with coronary artery (CA) ischemia, due to thrombosis of the native aortic root (NAR) and subsequent thromboembolic (TE) myocardial infarction (MI).

CASE REPORTS

Case 1

An 18-year-old male with HLHS with mitral stenosis (MS) and aortic atresia (AA) was initially palliated with a Stage I Norwood with modified Blalock–Taussig shunt followed shortly by balloon angioplasty of the reconstructed aortic arch. He subsequently underwent bidirectional Glenn (BDG) at 6 months of age and nonfenestrated extracardiac Fontan (ECF) operation with a 16 mm Impra tube graft at 4 years of age and had remained on lisinopril, aldactone, and aspirin.

The patient presented acutely to his local hospital with new-onset chest pain and shortness of breath (SOB). He was noted to have elevated troponin I (Trop-I) of 2.8 ng/ml (upper limit of normal, 0.15 ng/ml). On admission to the tertiary care center (TCC), the electrocardiogram (EKG) was significant for possible right ventricular (RV) hypertrophy with RV strain, ST depression in the anterior precordial leads, and T wave abnormality throughout the inferior and precordial leads. Trop-I had increased to 14.67 ng/ml with rising blood lactate to 3.36 mmol/l and a basic natriuretic peptide (BNP) level of 217 pg/ml. The echocardiogram (Echo) showed new moderate-to-severe RV systolic dysfunction with significant apical hypokinesis. He was taken urgently for cardiac catheterization (CCath) for hemodynamic evaluation and coronary angiography.

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Address for correspondence: Dr. Bhavi Patel, Department of Pediatric Cardiac Critical Care and Cardiology, Nicklaus Children's Hospital, 3100 S. W. 62nd Avenue, Miami, FL 33155, USA. E-mail: bhaviapatel@gmail.com

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CCath showed severely diminished cardiac index (CI) of 1.3 L/min/m^2 with elevated RV end diastolic pressure and Fontan pressure. Angiography revealed a large filing defect in the posterior sinus of the hypoplastic NAR consistent with thrombus of undetermined age [Figure 1]. The right CA appeared large and unobstructed. The left main and left circumflex CA appeared unobstructed, but the proximal left anterior descending (LAD) tapered abruptly, which was concerning for TE. After the identification of the large thrombus in the NAR with probable partial embolism to the LAD, tissue plasminogen activator (TPA) was administered directly into the NAR in the hopes of reducing the TE burden to the LAD and preventing further emboli. Stent angioplasty of the LAD was not deemed feasible due to the hypoplasia of the native left CA system as well as the significant risk of dislodging the remainder of the large NAR thrombus.

The patient returned to the Cardiac Intensive Care Unit (CICU) requiring inotropic support due to hypotension and myocardial dysfunction. Systemic TPA therapy was initiated to reduce further TE burden. After the CCath procedure, the patient had an acute decompensation that began with neurological posturing and culminated in cardiopulmonary arrest. A full resuscitation was undertaken, but return of spontaneous circulation (ROSC) could not be achieved and the patient expired.

Case 2

A 17-year-old female with HLHS with mitral atresia (MA) and AA and coarctation of the aorta (CoA) underwent Stage I at birth with balloon angioplasty of the CoA at 3 months of age. BDG was performed at 4 months of age followed by ECF with 5 mm fenestration at 4 years of age and subsequent closure of fenestration at 5 years of age. She was maintained on aspirin, lasix, amlodipine, lisinopril, aldactone, and carvedilol. She had moderately depressed ventricular function on Echo since her ECF operation.

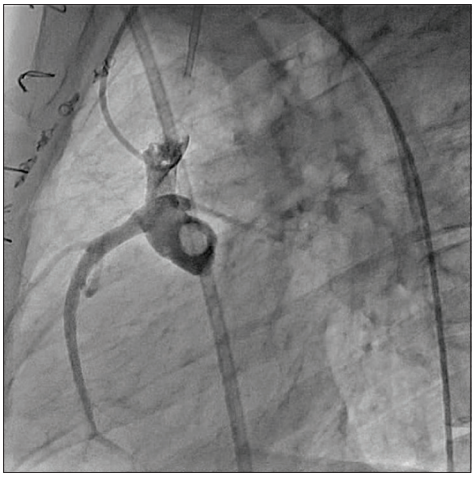

Figure 1: Angiography showing a large filing defect in the posterior sinus of the hypoplastic native aortic root consistent with thrombus

She presented to the local hospital with cough, nasal congestion, and subjective fevers for 2–3 days followed by the onset of intermittent, stabbing chest pain of 1-day duration. EKG showed depressed ST segments, and she had an elevated Trop-I. She was started on oxygen via nasal cannula and milrinone infusion and was transferred to a TCC.

On arrival to the TCC, she had stable hemodynamics with a BNP of 80 pg/ml and an elevated Trop-I of 45 ng/ml with normal lactate. Echo showed severely depressed RV function with anterior wall akinesia from the tricuspid valve to mid septum and dyskinesia of the RV apex. Computerized tomography angiography (CTA) of the heart showed thrombosis of the native ascending aorta extending to the right main CA, with opacification of two acute marginal branches distal to the thrombus, dilated neo-aortic root and ascending aorta, and no evidence of CoA. At this point, she was initiated on a heparin drip, Plavix, and continued on aspirin.

Within hours of starting anticoagulation, she had a V-Tach cardiac arrest requiring full resuscitation with ROSC; she was intubated and started on inotropes and lidocaine drips. After stabilization, she was transferred to a facility with heart-transplant capabilities. She was noted to have a left middle cerebral artery thrombotic stroke with right-sided weakness, expressive aphasia, and apraxia. She was treated with anticoagulation with heparin drip with recent Echo showing good flow in the CA and no thrombus in the NAR with depressed function. Currently, she is recovering at a chronic care facility with physical and occupational therapy.

Case 3

A 13-year-old male with a diagnosis of HLHS with MS and AA initially underwent Stage I palliation at 10 days of life, followed by BDG at 6 months of age and a 19-mm nonfenestrated ECF at 3 years of age, had remained on aspirin, but had been lost to follow-up for 10 years. Three weeks before admission, he started experiencing abdominal pain and SOB, for which he sought medical evaluation at an outside facility. He was transferred to a TCC for further evaluation and assessment.

On admission to the CICU, EKG demonstrated first-degree heart block with ST segment depression. Echo showed severely depressed ventricular function. He was initiated on oxygen via nasal cannula and milrinone infusion. Laboratories were significant for a hemoglobin of 18 g/dL, blood urea nitrogen of 26 mg/dL, creatinine of 1.27 mg/dL, alanine aminotransferase of 100 IU/L, aspartate aminotransferase of 182 IU/L, BNP of 1233 pg/ml, Trop-I of 15.82 ng/ml, and lactate of 2.35 mmol/L. During the first 6 h of admission to the CICU, he started experiencing chest pain associated with ST segment changes, a Trop-I elevation of 21 ng/ml, and creatine kinase MB of 140 ng/ml. He was emergently

taken for CCath where he was found to have CI of 1.1 L/min/m^2 and a Fontan pressure of 18 mmHg; a large thrombus in the LAD as well as thrombus formation within the NAR was visualized by angiography. After consultation with adult interventional cardiology, catheter intervention for the LAD thrombus was not recommended and the patient was started on heparin therapy. He was transferred to a cardiac transplant center where he is currently awaiting transplant.

DISCUSSION

Thrombosis and TE complications are well described in patients after the ECF operation; however, NAR thrombosis after the ECF operation in HLHS is very uncommon.[1-3] NAR thrombosis and TE phenomenon have been described in case reports of HLHS after Stage I palliation, BDG, and Fontan surgeries; however, this is one of the first case series of an embolic MI in patients with ECF circulation with an underlying diagnosis of HLHS with 10 years or more after the ECF operation. There have only been 13 reported cases of NAR thrombosis in HLHS, of which only four occurred after the Fontan operation.[4-8]

In a postmortem study of 122 patients who underwent the Norwood procedure, CA perfusion impairment was found in 27%, making it the most important cause of death in this series.[9] The majority of the patients had stenosis at the orifice of the aortic root coming off the neo-ascending aorta. Thrombosis within the NAR likely occurs due to flow stasis, which may be more prevalent in patients with AA without any forward flow, especially with native aortas of relatively large diameters. This statement i believe is appropriate and this is what it is trying to state. Flow stasis seen in native aortic root especially in lesions with aortic atresia leads to thrombus formation. Hence minimizing the length of the native aortic root by constructing the neo-aorta as proximal as possible with compromising the coronary ostia will limit the area of flow stasis and risk of thrombus formation. Also meticulous suturing to lower the risk of stenosis of the root orifice will also limit flow stasis and thus thrombus formation.[5,9] Common manifestations of root thrombosis in single ventricle patients include conduction disturbances, congestive heart failure, and systemic and coronary TE; and hypothesized risk factors include anatomic native valve abnormality, prolonged hospitalization, absence of anticoagulation therapy, length of the remnant NAR, and number of sutures.[6]

Currently, management decisions are made on a case-by-case basis and include anticoagulation alone, systemic thrombolysis, directed thrombolysis injected in the aortic root, and surgical thromboembolectomy. From our own experience, systemic and direct thrombolysis may carry a prohibitively high risk of stroke, either due to TE or primary intracranial hemorrhage. Although more data are needed to make strong recommendations, patients with HLHS, AA, and a relatively long native ascending aorta may benefit from long-term warfarin to prevent thrombus formation in the NAR.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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