Surgical Thrombectomy of Two Left Ventricular Thrombi in a Child With Acute Myocarditis

abstract

Myocarditis is a potentially life-threatening disease. Although ventricular thrombus formation in myocarditis is rare, it carries the risk of serious complications. We report on a 10-year-old previously healthy girl presenting with 2 large left ventricular thrombi in acute lymphocytic acute myocarditis. No coagulation disorder was found. Her clinical course and mobile thrombi characteristics prompted us to carry out an urgent surgical thrombectomy rather than primary anticoagulation therapy. The patient has recovered well without clinical signs of thromboembolism. Pediatrics 2013;131:e1–e5
Left ventricular thrombus formation is a serious complication in a variety of underlying diseases. Predisposing conditions include reduced myocardial contractility and left ventricular dilatation as in dilatative cardiomyopathy, myocardial infarction, and in very rare cases, myocarditis.1–5 Additionally, coexisting hematologic and coagulation disturbances may aggravate thrombus formation.4,5 Intracardiac thrombi are rare in children, but reports of their incidence have increased in the last few decades, especially in patients with hypercoagulation.6 When present, thrombi are a critical component in significant morbidity and mortality.7 We report on a 10-year-old girl with myocarditis who developed both a highly mobile mural and a pedunculated thrombus in the left ventricular apex.

**PATIENT PRESENTATION**

We report on an obese 10-year-old girl (BMI = 33) in good health. The family history was only remarkable in that the father has arterial hypertension. Two weeks before admission to our hospital, she presented with symptoms of an upper airway infection. She gradually developed dyspnea, fever, fatigue, and tachycardia, had an intermittent oxygen demand, and had abnormal laboratory tests under antibiotic therapy. On admission after transfer from a peripheral hospital, she was in stable but reduced general condition. Her blood pressure was 92/48 (58) mm Hg, with a heart rate of 130 beats per minute. We noted a gallop rhythm with no murmur. In addition, orthopnea was present at a breathing rate of 35 breaths per minute, with a 96% saturation rate without oxygen. A chest radiograph revealed evidence of a slightly increased heart/chest ratio of 0.51, minor central venous congestion, and small bilateral pleural effusions. Echocardiography demonstrated a dilated left ventricle, globally reduced left ventricular contractility with an ejection fraction of 46%, and a shortening fraction of 25%, with no signs of mitral or tricuspid valve regurgitation. No thrombus was visible at that time. Electrocardiogram findings and relevant laboratory tests are shown on Fig 1 and Table 1.

Along with bed rest, we initiated anti-congestive medical therapy with a β-blocker, diuretics, and low-dose therapy with low-molecular-weight heparin. We gradually observed an improvement in her clinical status, laboratory values, and left ventricular contractility, but between 48 and 72 hours after admission, we observed a hyperintense mural area in the left ventricular apex without any regional hypocontractility in the area of the thrombus (Table 1). This area developed rapidly to 1 pedunculated (14 × 18 mm, with a stalk of 5 mm) and 1 highly mobile thrombus (15 × 20 mm) clearly identified in 2-dimensional echocardiography (Fig 2 and Supplemental Movie). To prevent thromboembolism, surgical thrombectomy was urgently initiated on the fourth day after admission. The dimensions of the thrombi compared with the aortic valve (17 mm in diameter) and truncus bicaroticus (16 mm at its origin) had a major impact on our decision.

Using mild hypothermic cardiopulmonary bypass and induced ventricular fibrillation, we incised the left ventricular apex. The ventricular myocardium appeared normal macroscopically, and there was no evidence of noncompaction or hypertrabeculation that could offer an exposed area for thrombus formation. A white, tender, pedunculated thrombus and highly mobile septal thrombus were removed (Fig 2). Moreover, endomyocardial biopsies were taken from the left ventricular apex. Histologic examination confirmed the diagnosis of myocarditis via lymphocyte-rich infiltration of the myocardium. The surgical and postoperative courses were uneventful. Despite a ventriculotomy in an inflamed myocardium, weaning from cardiopulmonary bypass proceeded satisfactorily under medical treatment with adrenaline at a maximum dose of 0.15 µg/kg/min and milrinone at a dose of 0.7 µg/kg/min. We could reduce all inotropes rapidly. She was weaned quickly from ventilation, and extubation occurred 5 hours after surgery. She showed no clinical signs of any neurologic impairment or other symptoms of thromboembolism. Immediately after the operation, anticoagulation therapy with intravenous unfractioned heparin was started. The partial thromboplastin time level was maintained at between 50 and 80 seconds. On postoperative day 4, anticoagulation was switched to subcutaneous low-molecular-weight heparin; the anti-factor Xa activity level was maintained at between 0.5 and 0.8 IU/mL to prevent any new thrombus formation. Interestingly, the D-dimer level rose after surgery to a maximum of 10 mg/L, which we attributed to the fibrinolytic activity of heparin rather than new thrombus formation. This attribution was supported by the slow drop in the D-dimer value over time (Table 1). The patient was discharged on day 17 after thrombectomy with normal left ventricular contractility, no thrombus formation, and a normal neurologic examination. The extensive laboratory workup of etiologic factors was all normal, including thrombophilic factors. Thrombophilia testing included protein S deficiency, protein C deficiency, activated protein C resistance, antithrombin deficiency, Factor V Leiden mutation, prothrombin 20210 mutation, Factor VIII, lupus anticoagulant, and homocysteine. In the follow-up over 4 months, the patient remained well; she presented no signs of any new thrombus formation.

**DISCUSSION**

Acute myocarditis is a potentially life-threatening disease. Etiologic factors
are usually virus infections, but immune-mediated diseases including acute rheumatic fever, collagen vascular diseases, and toxic agents may also be a cause. Acute myocarditis can lead to left ventricular systolic dysfunction and dilatation. Ventricular dysfunction in turn is associated with endothelial function abnormalities, which may give rise to thrombus formation caused by endocardial injury, the procoagulant effects of cytokines, and blood flow stasis. In addition, hypercoagulation (eg, caused by protein C and protein S deficiencies) may contribute to thrombus formation. Although ventricular thrombus formation is rare in myocarditis, it does carry the risk of complications by thromboembolism and by impairing ventricular function.

Controversy remains as to therapy recommendations, and the optimal treatment of pediatric patients with intracardiac thrombosis should be specified. The traditional therapy for intracardiac thrombi in pediatric patients has been surgical thrombectomy, but there are increasing reports of non-surgical strategies. A single-center study demonstrated medical therapy alone as an effective alternative to surgical thrombectomy in pediatric patients, including those after the Fontan procedure. Conversely, additional key findings are significant morbidity secondary to thromboembolism and a poorer prognosis with left ventricular thrombus.

### Table 1

<table>
<thead>
<tr>
<th></th>
<th>Admission</th>
<th>Day -2/1</th>
<th>Surgery</th>
<th>Day +1</th>
<th>Day +5</th>
<th>Discharge</th>
</tr>
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<tbody>
<tr>
<td>Troponin T (ng/mL)</td>
<td>1.65</td>
<td>—</td>
<td>0.17</td>
<td>—</td>
<td>—</td>
<td>0.021</td>
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<tr>
<td>Creatinin kinase (U/L)</td>
<td>130</td>
<td>—</td>
<td>45</td>
<td>397</td>
<td>27</td>
<td>29</td>
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<tr>
<td>ProBNP (pg/mL)</td>
<td>16 190</td>
<td>—</td>
<td>4749</td>
<td>—</td>
<td>1950</td>
<td>2159</td>
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<tr>
<td>CRP (mg/L)</td>
<td>94</td>
<td>41</td>
<td>24</td>
<td>62</td>
<td>49</td>
<td>17</td>
</tr>
<tr>
<td>D-dimeres (mg/L)</td>
<td>7.06</td>
<td>—</td>
<td>4.19</td>
<td>1.74</td>
<td>10</td>
<td>1.97</td>
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<tr>
<td>Fibrinogen (mg/dL)</td>
<td>525</td>
<td>—</td>
<td>483</td>
<td>357</td>
<td>836</td>
<td>406</td>
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<tr>
<td>Anti-Xa level (U/L)</td>
<td>—</td>
<td>—</td>
<td>0.21</td>
<td>0.15</td>
<td>0.37</td>
<td>0.77</td>
</tr>
<tr>
<td>Platelets (G/L)</td>
<td>316</td>
<td>351</td>
<td>412</td>
<td>233</td>
<td>429</td>
<td>408</td>
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<tr>
<td>Leukocytes (G/L)</td>
<td>17.8</td>
<td>10.7</td>
<td>10.1</td>
<td>15.8</td>
<td>13.3</td>
<td>9.8</td>
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<tr>
<td>EF (%)</td>
<td>46</td>
<td>48</td>
<td>50</td>
<td>—</td>
<td>55</td>
<td>62</td>
</tr>
<tr>
<td>SF (%)</td>
<td>23</td>
<td>24</td>
<td>25</td>
<td>—</td>
<td>28</td>
<td>33</td>
</tr>
</tbody>
</table>

Day -2/1 represents the time between the second and third days after admission. Anti-Xa, anti-factor Xa activity; CRP, C-reactive protein; EF, ejection fraction; ProBNP, Pro brain natriuretic peptide; SF, shortening fraction; —, no measurement.
To ensure adequate therapy, we find it important to differentiate thrombus characteristics. Mural thrombi normally have a broad fixed base on the cardiac wall and show a minor tendency to cause thromboembolism. Mobile or pedunculated thrombi are rarer than mural thrombi but are believed to increase the risk for thromboembolism considerably. This thesis is supported by echocardiographic studies addressing the embolic potential of left ventricular thrombi, which have identified thrombus protrusion and mobility as high-risk factors for subsequent embolism. Additionally, larger intracardiac thrombi tend to embolize more frequently, and the development of a cerebral embolism is a serious complication. There are few studies on the incidence of thromboembolism in children with intracardiac thrombi, but 2 studies with a relatively large cohort reported an incidence of 13% to 14%, whereas autopsy studies described an even higher frequency.

Under anticoagulation therapy, a pedunculated thrombus may become unstable because of additional narrowing of the thrombus stalk, thus promoting embolism. Concerning the dynamics of thrombus development under prophylactic anticoagulation in our patient, the time required to reduce the thrombus size via medical therapy alone may well have been too long and thus have put our patient at an additional risk for thromboembolism. Because of the thrombi dimensions, the truncus bicuspidicus was prone to occlude in thromboembolism, possibly causing a fatal outcome.

Conversely, underlying myocarditis may limit surgical options. Generally speaking, the risk of mortality after surgery depends on the extent of pre-operative left ventricular dysfunction. One would anticipate additional deterioration in ventricular function by cardiopulmonary bypass and cardioplegic arrest in myocarditis. We decided on immediate surgery because the patient's ventricular function had already started to improve again (Table 1). Thus, we assessed the risk of a potentially life-threatening thromboembolism during anticoagulation treatment to be greater than the risk of surgery-induced deterioration in myocardial function. There are different ways to surgically access the thrombi: aortotomy, atrial access, and ventriculotomy. We did not consider the aortic route because this would require cardioplegic arrest and impaired visibility in the apical region. Taking the atrial route might be less invasive than ventriculotomy, because no incision to the left ventricle is needed. Atrial access without cardioplegia is another
possibility but carries the potential of incomplete removal with reduced visibility. On the other hand, via induced ventricular fibrillation and limited ventriculotomy, one can safely explore the left ventricular cavity without cardioplegia. The main reason we preferred ventriculotomy to aortic or atrial access was to have two advantages: no need for cardioplegia in myocarditis and secure access to the thrombi.

We detected no infectious or coagulation abnormalities. During thrombus formation, left ventricular contractility was already improving and was better than in previously reported cases of ventricular thrombus formation. A potential explanation is that the acute infection itself had provoked hypercoagulation, inducing thrombus formation in already-improving left ventricular function. Additionally, the prophylactic use of low-molecular-weight heparin might not be adequately effective in an induced hypercoagulable state caused by myocarditis.

CONCLUSIONS

We describe a patient with rapid left ventricular thrombus formation during myocarditis. Notably, the left ventricular contractility was already improving at the time of supposed thrombus formation, and no promoting conditions were detectable. We considered surgical thrombectomy to be superior to anticoagulation therapy alone in light of the particular characteristics of these thrombi. Whether more intensive anticoagulation therapy can prevent thrombus formation will have to be elucidated; it may be worth consideration in infectious myocarditis. In addition, we advise frequent echocardiographic monitoring in myocarditis despite improving ventricular contractility.

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REFERENCES

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