Paediatric Takayasu’s arteritis complicated by thrombotic occlusion of the distal thoracic aorta

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Abstract

We present the case of a 1-year-old girl with mid-aortic syndrome due to untreated Takayasu’s arteritis who developed cardiogenic shock. Enhanced computed tomography revealed long-segment occlusion of the distal thoracic aorta. We successfully performed graft interpose (10 mm in diameter) under cardiopulmonary bypass through both median sternotomy and left posterolateral thoracotomy. The thrombus was relatively small and the distal thoracic aorta was narrow over a long segment due to severely thickened intima. Follow-up computed tomography showed widely patent graft without a stenotic region in the abdominal aorta or its branches. The patient discharged ambulatory without major complications.

Keywords: Pediatric Takayasu’s arteritis • Thrombotic occlusion • Mid-aortic syndrome

INTRODUCTION

Takayasu’s arteritis (TA) is an inflammatory, granulomatous and fibrosing arteritis of the aorta and its major branches [1, 2]. Paediatric TA is a devastating disease with significant morbidity and mortality. However, the diagnosis of paediatric TA might be difficult, partly because the clinical features of TA are non-specific [1]. We encountered a 1-year-old girl with untreated TA who developed cardiogenic shock due to thrombotic occlusion of the distal thoracic aorta.

CASE REPORT

A 1-year-old girl (height, 80 cm; weight, 10 kg) was referred to our institution due to cardiogenic shock. Chest X-ray showed cardiomegaly and severe lung congestion. PaO₂/FiO₂ ratio was 127. Echocardiogram revealed reduced ejection fraction and moderate mitral regurgitation without intracardiac or coronary anomalies. Colour Doppler showed obstruction in the arch distal to the left subclavian artery. Enhanced computed tomography (CT) revealed long-segment obstruction from the distal arch to the left subclavian artery. Enhanced computed tomography showed long-segment occlusion of the distal thoracic aorta. We successfully performed graft interpose (10 mm in diameter) under cardiopulmonary bypass through both median sternotomy and left posterolateral thoracotomy. The thrombus was relatively small and the distal thoracic aorta was narrow over a long segment due to severely thickened intima. Follow-up computed tomography showed widely patent graft without a stenotic region in the abdominal aorta or its branches. The patient discharged ambulatory without major complications.

Postoperatively, the patient suffered from acute renal failure and peritoneal dialysis was performed until postoperative Day 18. Histopathologic findings revealed fibrous thickening of the aorta and destruction of the arterial medial elastic fibres by lymphocytes and granulocytes. Therefore, mid-aortic syndrome due to TA was the definitive diagnosis. Enalapril, steroid and aspirin were initiated. Postoperative echocardiography demonstrated good left ventricular function without mitral regurgitation. Postoperative CT showed widely patent graft (Fig. 2). There was no stenotic region in the abdominal aorta or its branches. She was discharged ambulatory without major complications on postoperative Day 59.
Surgical interventions for TA have been performed to repair atypical aortic coarctation or involvement of abdominal arteries [1, 2]. Most such cases were diagnosed with TA preoperatively and treated with steroids and/or immunosuppressive agents. Moreover, patients with active inflammation at the time of surgery may be at higher risk for vascular complication [2] and controlling the inflammatory process before surgery is preferable.

Mid-aortic syndrome is characterized by a diffuse narrowing of the distal thoracic or abdominal aorta, regardless of aetiology. Percutaneous intervention is employed at many institutions to relieve the stenosis with good results [3]. However, endovascular therapy was considered too risky in this patient, because of the preoperative shock status and the long-segment occlusion.

If collateral circulation restores blood flow to the abdomen in a patient with long-segment aortic occlusion, scheduled revascularization without cardiopulmonary bypass can be performed [4]. Alternatively, if the patient has a bigger physique, a salvage operation such as axillo-external iliac artery bypass can be performed [5]. However, in our patient, we had no choice but to perform revascularization by graft interpose (it was impossible to perform arch reconstruction with native tissue because of long-segment obstruction and the severely thickened intima). Judging from her preoperative heart and lung condition, it appeared dangerous to perform revascularization only through a thoracotomy. Thus, we performed a sternotomy to place her on cardiopulmonary bypass promptly and enable central extracorporeal membrane oxygenation support in case of postoperative cardiac or respiratory failure.

One may argue that extra-anatomic ascending aorta to abdominal aorta bypass might be easily performed via median sternotomy and split midline laparotomy [4]. However, long and curved graft for such a small child should be avoided, because it is difficult to predict how the extra-anatomic graft will be stretched as the child grows, and such a graft would make repeated procedures necessary as the child grows.

In general, some patients with TA will experience a flare up. Patients with lower body mass index have a high risk of poor outcomes [1]. Moreover, disease activity of TA can increase the likelihood of graft revision [2]. Therefore, close monitoring, drug management and long-term follow-up are required. In order to detect new vascular complications early, it is necessary to perform enhanced CT on a regular basis.

**Conflict of interest:** none declared.
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