Paediatric cardiac surgery in a patient with cold agglutinins

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Abstract

Cold agglutinins (CAs) lead to organ thrombosis or haemolysis due to increased blood viscosity and red blood cell clumping when blood temperature drops below the thermal amplitude for haemagglutination. Although it is well known that CAs are particularly relevant to adult cardiac surgery with hypothermic cardiopulmonary bypass (CPB), paediatric cardiac surgery with congenital heart disease and with CAs has been reported very rarely. We present here a case of paediatric cardiac surgery to repair atrial septal defect with pulmonary stenosis in an 11-month-old infant with a family history of CAs. She was detected to have a high titre of CAs preoperatively, and underwent an intracardiac repair with normothermic CPB using temporary electrical fibrillation for added safety. Her post-operative course was uneventful without any complications.

Keywords: Cold agglutinins • Family history • Paediatric cardiac surgery • Normothermia • Electrical fibrillation

INTRODUCTION

Cold agglutinins (CAs) are autoantibodies that lead to haemagglutination and microvascular thrombosis at low temperature, followed by complement fixation and haemolysis during rewarming. The phenomenon appears sometimes in cardiac surgery with hypothermic cardiopulmonary bypass (CPB) and cardioplegia in patients with activated CAs. The incidence of CA-related complications during cardiac surgery is reported to be ~0.8% [1]. In this study, we present a case of paediatric cardiac surgery performed in an infant who was detected to have CAs preoperatively.

CASE REPORT

An 11-month-old infant weighing 7.7 kg was admitted to our hospital for surgical treatment of congenital heart disease. At 3 months after birth, her family physician found that she had heart murmur, and diagnosed her with atrial septal defect (ASD) and pulmonary stenosis (PS). Physical examination showed a systolic ejection murmur III/VI and fixed splitting of the second heart sound at the left second intercostal space. Echocardiogram examination revealed secundum ASD (12 × 8 mm) with left-to-right shunt, and valvular and supravalvular PS. Doppler examination demonstrated a velocity of 3.8 m/s across the pulmonary valve, corresponding to an instantaneous peak systolic pressure gradient of 58 mmHg. Cardiac catheterization for percutaneous balloon pulmonary valvuloplasty had been performed at 6 months after birth. Because it was effective against valvular PS but not supravalvular PS, the patient was considered a candidate for surgical repairs of the ASD and supravalvular PS. On admission, the preoperative blood testing demonstrated an elevated CA titre at 4°C (1:512) and at 25°C (1:64). The coagulation profile was normal. Family history revealed that her mother also suffered from ASD and CAs. Because we did not have further information of CAs, including the exact temperature below which haemagglutination due to CA activation occurs, open heart surgery with normothermic CPB using electrical fibrillation was planned for added safety.

The operation was performed via standard median sternotomy under general anaesthesia. The arterial cannula was inserted in the ascending aorta, and venous return was through bicaval cannulation. Normothermic CPB was initiated and maintained at 35.8°C at the lowest rectal temperature. The ASD was closed with an autologous pericardial patch under temporary electrical fibrillation. After the ASD closure, the heart was defibrillated. The supravalvular PS was released by pulmonary artery patch plasty with autologous pericardium on the subsequent normothermic CPB. The CPB time and the electrical fibrillation time were 95 and 31 min. No blood products were required in this operation. The patient was extubated in the operation theatre and stayed one night in the intensive care unit. No evidence of CA-related complications, such as microvascular thrombosis, cerebral infarction/bleeding, myocardial infarction or renal/hepatic insufficiency, were observed perioperatively. Serum concentrations of both cardiac troponin T (cTnT) and heart fatty acid-binding protein (HFABP), as specific markers for perioperative myocardial damage [2], were low at 1 h after operation (cTnT 3.74 ng/ml, HFABP 53 ng/ml). The echocardiograms taken immediately and at 7 days after surgery showed a good wall motion of the left ventricle with an improvement in the supravalvular PS and no residual ASD. The patient was discharged at 8 days after operation uneventfully.
Although there are studies reporting the perioperative risks of cardiac surgery in a patient with CAs and the need for surgical techniques with regard to CPB and myocardial protection, paediatric cardiac surgery in children with congenital heart disease and CAs has been reported very rarely. Generally, perioperative management of cardiac surgery in patients with CAs depends on the CA titre and the temperature below which CA activation occurs. Patients with high CA titre and high thermal amplitude require special management according to the surgical procedures, whereas patients with low CA titre and low thermal amplitude undergo cardiac surgery with routine management. To avoid CA activation, normothermic CPB with varying techniques of myocardial protection, including warm cardioplegia, intermittent cross-clamping or induced ventricular fibrillation, has been performed in adult patients previously [1, 3–5]. Agarwal et al. reported 13 adult patients with high CA titre who underwent cardiac surgery including coronary artery bypass grafting, mitral valve replacement and ASD closure. They had a thermal amplitude of 4–22°C, which mean that they had no risk for mild hypothermic CPB. However, agglutination could have developed in coronary arteries with the use of cold cardioplegia at 4°C. Avoidance of cold cardioplegia should be necessary for those patients. In the present case, the patient had a positive family history of ASD and CAs. Her mother had been detected with a high titre of CAs when she underwent cardiac surgery (ASD closure) in the childhood, but the details of the surgical techniques and outcomes were unknown. The preoperative blood testing of the patient showed an elevated CA titre at 4 and 25°C, but we did not get any further indications of CAs including the exact temperature below which haemagglutination due to CA activation occurs. To avoid CA activation due to mild hypothermic CPB and cold cardioplegia, paediatric cardiac surgery with normothermic CPB using temporary electrical fibrillation was selected as added safety in this case.

In our institution, normothermic CPB with electrical fibrillation is frequently used for isolated ASD repair through a right posterolateral thoracotomy in female children [6]. Although cardioplegic arrest with aortic clamping is possible in the present case, we selected the normothermic CPB with electrical fibrillation because its safety is well established when the electrical fibrillation is performed in a short period with acceptable perfusion pressures. In the present case, no perioperative myocardial damage was detected by using cTnT and HFBP, both of which are early specific markers for assessment of myocardial damage in paediatric cardiac surgery [2].

In summary, we experienced a case of paediatric cardiac surgery (ASD and PS repairs) in a patient with CAs. Normothermic CPB with electrical fibrillation can be one of the safe surgical techniques with a successful outcome for patients with CAs. We recommend preoperative evaluation of CA titre and temperature in the CA activation, which can give us an individualized surgical planning to prevent CA-related complications.

**Conflict of interest:** none declared.

**REFERENCES**