A rare cause of recurrent modified Blalock-Taussig shunt thrombosis: Antiphospholipid antibodies

Neslihan Kplaplar, Ersin Errek*, Ender Ödemis, Erkut Öztürk
Department of Pediatric Cardiology and Pediatric Cardiovascular Surgery, Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital; Istanbul-Turkey

Introduction

The modified Blalock-Taussig (mBT) shunt is a palliative surgical treatment that increase pulmonary blood flow in patients with cyanotic congenital heart diseases. The incidence of mBT shunt thrombosis is reported to be among 1-17% (1). Several risk factors were defined about shunt thrombosis such as small age, low body weight, hypoplastic pulmonary arteries, small graft size and thrombophilia (1). We present a case of recurrent mBT shunt thrombosis associated with elevations in antiphospholipin antibody levels.

Case Report

A 22 month-old boy was admitted to our clinic with cyanosis. Arterial oxygen saturation (SpO₂) was measured 50%. A total blood count, partial thromboplastin time, prothrombin time, biochemical parameters were normal. Transthoracic echocardiography (ECHO) demonstrated that transposition of great arteries, L-malposition of great arteries, ventricular septal defect (outlet), severe pulmonary stenosis (valvular, subvalvular). Systolic gradient between pulmonary artery and left ventricle was 90 mm Hg. Both right and left pulmonary artery was measured 6 mm. A right mBT shunt which using a polytetrafluoroethylene tube graft 5 mm in diameter was performed. After surgery we administered heparin infusion of 10 IU/kg/h and oral aspirin 5 mg/kg/day to prevent graft thrombosis. Several hours after surgery, his SpO₂ suddenly dropped below 50% and ECHO showed thrombotic occlusion of the shunt. He underwent angiography and first, heparin was given into the shunt, after then streptokinase and tissue plasminogen activator was administered. After shunt was successfully recanalyzed, immediately re-occluded and we performed balloon angioplasty promptly. Shunt was thrombosed repeatedly. Although stent placement into the graft, shunt flow remained insufficient. Therefore recanalization of the shunt with the same sized graft was performed surgically. Postoperatively shunt flow was inadequate and after heparin bolus was started, shunt flow increased dramatically. Because of this clinical picture, we suggest that thrombophilia and protein C, protein S, antithrombin III, prothrombin G20210A mutation, factor V Leiden mutation, folic acid, vitamin B12, homocysteine levels were measured. All of them were normal. In addition, we measured to antiphospholipid antibody (APA) levels and lupus anticoagulant and antiphospholipin antibodies were positive. We administered low-molecular-weight heparin and aspirin. He discharged with these therapy after five days and control laboratory tests showed that an undetectable level of APA. He was unremarkable during 13 months.

Discussion

Traditionally, diagnostic criteria of antiphospholipid syndrome (APS) in children was defined as: A combination of two clinical (thrombosis/recurrent abortions) and one of three laboratory features which are positive APA (lupus anticoagulant or antiphospholipin or anti-β2GPI) present on two occasions at minimum 12 weeks apart (2). However currently there is a quite controversy about diagnostic criteria of APS. Presence of antiphospholipin antibodies detect several other conditions, such as infants with atop dermatitis, juvenile idiopathic arthritis, infections and vaccinations. It has been reported in healthy children also (3). Furthermore some reports were published on seronegative patients who have the same clinical features with seropositive patients in APS (4, 5). Moreover there is a large variation about standardization and methods of measurement in APA levels. For these rea-
sons, it is unclear how interpret APA levels in APS. Also Rodriguez-Garcia et al. (5) showed that there is no differences between seronegative and seropositive APS in terms of thrombotic events. A case report which was published by Middlebrooks et al. (6) there is an association between temporary APA positivity and recurrent stent thrombosis. This case was explained by seronegative APS or long-term antithrombotic therapy by the authors. Additionally it was shown that especially aspirin can reduce APA levels (7). In our patient, there is no other possible risk factors responsible for shunt thrombosis except positive APA levels. Based on publications mentioned above we affirm that mBT shunt thrombosis might be related to positive APA levels in this case.

Certain thromboembolic factors such as protein C deficiency, factor V Leiden or protombin 20210 mutation have been reported with cardiac thrombosis in children (8, 9). Deally et al. (10) published a newborn with mBT shunt thrombosis caused by APS in 1999. This case has antithrombin III deficiency, also. However, there was only one trombophilic factor in our patient. He is the first case in the literature in this regard.

Conclusion

Especially in patients with recurrent shunt thrombosis, hereditary thrombophilia should be investigated and APA levels should be measured in these patients.

References