evidence to the substantial literature on usefulness of this assessment in diagnosis of complex anatomical pathologies of the heart.

Video 1. 2-D transthoracic apical 4-chamber view showing the systolic anterior motion of the mitral valve
Video 2. Three dimensional TEE image showing the anatomic relation of the ventricular septal aneurysm (Asterisk) to Aorta (AO), Left Atrium (LA), Right Ventricle (RV) and Tricuspid Valve (TV)
Video 3. Ventriculography in left cranial projection showing VSA located just beneath the aortic valve

References

1. Choi M, Jung JI, Lee BY, Kim HR. Ventricular septal aneurysms in adults: findings of cardiac CT images and correlation with clinical features. Acta Radiol 2011; 52: 619-23. [CrossRef]
2. Panduranga P, Al-Riyami AA. Coronary anomaly with a left ventricular accessory chamber: Is it left ventricular aneurysm or diverticulum? J Saudi Heart Assoc 2012; 24: 191-4. [CrossRef]
3. Jang SW, Rho TH, Kim JH. Membranous interventricular septal aneurysm resulted in complete atrioventricular block. Heart 2010; 96: 244. [CrossRef]
4. Graffigna A, Minzioni G, Ressia L, Vigano M. Surgical ablation of ventricular tachycardia secondary to congenital ventricular septal aneurysm. Ann Thorac Surg 1994; 57: 921-4. [CrossRef]
5. Edelstein J, Charms BL. Ventricular septal aneurysms. A report of two cases. Circulation 1965; 32: 981-4. [CrossRef]
6. Yamauchi H, Matsui Y, Shiiya N, Murashita T, Sakuma M, Yasuda K. Dissection of the interventricular septum with aorto-left ventricular communication due to infective endocarditis: report of a rare case. Nihon Kyobu Geka Gakkai Zasshi 1996; 44: 534-8.
7. Mariscalco G, Blanzola C, Leva C, Cattaneo P, Mantovani V, Ferrarese S, et al. Unruptured ventricular septal wall dissection. A case report. J Cardiovasc Surg (Torino) 2006; 47: 349-52.
8. Kaplan M, Demirtaş M, Sayrak H, Çimen S, Dağsalı S, Özler A. An anatomopathologic study of membranous septum aneurysms and significance of their surgical treatment. Cardiovasc Surg 2000; 8: 561-6. [CrossRef]
9. Chang CY, Hsiung MC, Tsai SK, Wei J, Ou CH, Chang YC, et al. Live three-dimensional transesophageal echocardiography in an unusual case of aorto-left ventricular tunnel with a large interventricular septal aneurysm. Echocardiography 2011; 28: E12-5. [CrossRef]
10. Afaneh AB, Wymer DC, Kraft S, Winchester DE. Membranous ventricular septal aneurysm diagnosed by means of cardiac computed tomography. Tex Heart Inst J 2012; 39: 450-1.

Address for Correspondence: Dr. Barış Güngör,
Mehmet Akif Mah. Hicret Sok. Merit Life Bulvar, A/3 Ümraniye, İstanbul-*Türkiye*
Phone: +90 216 545 86 95
Fax: +90 216 459 27 66
E-mail: drbarisgungor@gmail.com
Available Online Date: 09.06.2014

DOI:10.5152/akd.2014.5282

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

Neslihan Kıplapınar, Ersin Erek*, Ender Ödemiş, Erkut Öztürk
Department of Pediatric Cardiology and *Pediatric Cardiovascular Surgery, Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital; İstanbul-*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 anticardiolipin antibody levels.

Case Report

A 22 month-old boy was admitted to our clinic with cyanosis. Arterial oxygen saturation (SpO_2) 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-malpozition 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 SpO_2 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 anticardiolipin 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 one of two clinical (thrombosis/recurrent abortions) and one of three laboratory features which are positive APA (lupus anticoagulant or anticardiolipin or anti-β2GP1) present on two occasions at minimum 12 weeks apart (2). However currently there is a quite controversy about diagnostic criteria of APS. Presence of anticardiolipin antibodies detect several other conditions, such as infants with atopic 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 that 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 authours. 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 protrombin 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

1. Tsai KT, Chang CH, Lin PJ. Modified Blalock-Taussig shunt: statistical analysis of potential factors influencing shunt outcome. J Cardiovasc Surg (Torino) 1996; 37: 149-52.
2. Ravelli A, Martini A. Antiphospholipid antibody syndrome in pediatric patients. Rheum Dis Clin North Am 1997; 23: 657-76. [CrossRef]
3. Avcin T, Ambrozic A, Bozic B, Accetto M, Kveder T, Rozman B. Estimation of anticardiolipin antibodies, anti-beta2 glycoprotein I antibodies and lupus anticoagulant in a prospective longitudinal study of children with juvenile idiopathic arthritis. Clin Exp Rheumatol 2002; 20: 101-8.
4. Ruan Y, Bridges JS, Kumar K, Raphael JA, Acharjee S, Welty FK. Complete resolution of a mitral valve vegetation with anticoagulation in seronegative antiphospholipid syndrome. Clin Rheumatol 2008; 27: 1577-9. [CrossRef]
5. Rodriguez-Garcia JL, Bertolaccini ML, Cuadrado MJ, Sanna G, Ateka-Barrutia O, Khamashta MA. Clinical manifestations of antiphospholipid syndrome (APS) with and without antiphospholipid antibodies (the so-called 'seronegative APS'). Ann Rheum Dis 2012; 71: 242-4. [CrossRef]
6. Middlebrooks EH, Panda M. Multiple recurrent stent thrombosis in a patient with coexisting clopidogrel resistance and increased anticardiolipin antibodies: a case report. Case Rep Med 2010; 2010: 974149.
7. Ikonomidis I, Lekakis J, Vamvakou G, Loizou S, Revela I, Andreotti F, et al. Aspirin reduces anticardiolipin antibodies in patients with coronary artery disease. Eur J Clin Invest 2006; 36: 839-43. [CrossRef]
8. Watanabe M, Aoki M, Fujiwara T. Thrombotic occlusion of Blalock-Taussig shunt in a patient with unnoticed protein C deficiency. Gen Thorac Cardiovasc Surg 2008; 56: 544-6. [CrossRef]
9. Gürgey A, Özyürek E, Gümrük F, Çeliker A, Özkutlu S, Özer S, et al. Trombosis in children with cardiac pathology: frequency of factor V Leiden and prothrombin G20210A mutations. Pediatr Cardiol 2003; 24: 244-8. [CrossRef]
10. Deally C, Hancock BJ, Giddins N, Hawkins L, Odim J. Primary antiphospholipid syndrome: a cause of catastrophic shunt trombosis in the newborn. J Cardiovasc Surg (Torino) 1999; 40: 261-4.

Address for Correspondence: Dr. Neslihan Kıplapınar,
Mehmet Akif Ersoy Göğüs Kalp Damar Cerrahisi Eğitim ve Araştırma Hastanesi, İstasyon Mah. İstanbul Cad. Bezirganbahçe Mevki 34303 Küçükçekmece, İstanbul-*Türkiye*

Phone: +90 212 692 20 00-1446
Fax: +90 212 471 94 94
E-mail: nmelikoglu@yahoo.com

Available Online Date: 09.06.2014

DOI:10.5152/akd.2014.5279

A very rarely seen cardiac mass (Rosai-Dorfman disease)

Ersan Özbudak, Ali Ahmet Arıkan, Şadan Yavuz, Ümit Halıcı[1], Turan Berki
Department of Cardiovascular Surgery, Faculty of Medicine, Kocaeli University; Kocaeli-*Turkey*
[1]Clinic of Cardiovascular Surgery, Samsun Education and Research Hospital; Samsun-*Turkey*

Introduction

Sinus histiocytosis (Rosai-Dorfman Disease) is a rare disease, which is characterized by massive lympadenopathies with unknown etiology. It was first defined in 1969 by Rosai and Dorfman (1). Although it is seen most frequently in the first two decades of life, it can be observed at any age. The frequency of cardiac involvement is less than 1% in Rosai-Dorfman disease (RDD) (2). Here, we report a case with extranodal RDD in which cardiac involvement was detected.

Case Report

A 62-years-old male patient was referred to cardiology clinic with the complaints of atypical chest pain and dyspnea. His physical examination was unremarkable. A cardiac mass with 2x1.8 cm dimentions attached to wall of the right atrium was observed in transthoracic echocardiography (Fig. 1). Thoracic computerized tomography (CT) (Fig. 2) and Cardiac Magnetic Rezonans (MRI) showed a mass with 37x29 mm dimensions originating from the wall of the superior vena cava and extending to the interatrial septum, and along the lateral right atrial wall to the atrioventricular groove (Fig. 3A, B).

Surgical technique

Under general anesthesia, median sternotomy, standart aortic cannulation and selective bicaval cannulation were performed. A gray-

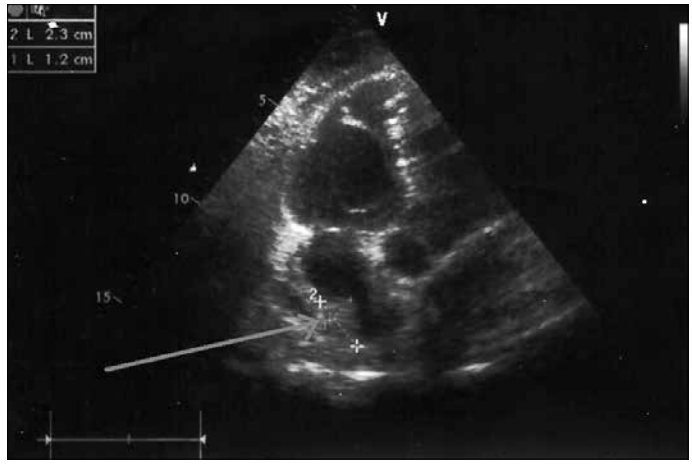

Figure 1. Showing a right atrial mass in Echocardiography