Pediatr Cardiol 13:173–175, 1992

Pediatric Cardiology

Development of a Left Atrial Ball Thrombus in a Woman with Complex Congenital Heart Disease Including Congenital Mitral Valve Stenosis

Mubadda Salim and Brian W. McCrindle

Department of Pediatrics and Division of Pediatric Cardiology, The Johns Hopkins University School of Medicine, Baltimore, Maryland, USA

SUMMARY. **A 20-year-old woman with complex cyanotic congenital heart disease, including severe congenital mitral stenosis and intact atrial septum, who developed a left atrial ball thrombus and an embolic phenomenon, is presented. Increased vigilance in this unique setting is necessary for the antemortem detection of this rare complication.**

KEY WORDS: **Left atrial thrombus—Congenital mitral valve stenosis—Congenital heart disease**

Left atrial thrombus in adults has been described as a complication of acquired obstructive mitral valve disease, usually in the setting of associated atrial tachyarrhythmias [7, 9]. Only rarely has left atrial thrombus been described in the context of congenital mitral valve obstruction [1, 5, 12]. Advances in echocardiography have improved the detection of intracardiac thrombi and have allowed antemortem treatment. This report describes a case of left atrial ball thrombus in a young adult patient with complex cyanotic congenital heart disease including the presence of severe congenital mitral valve stenosis and intact atrial septum.

Case Report

A 2.4-kg female infant was noted at birth to have multiple malformations including an atretic right auditory meatus and phocomelia of the upper extremities. Chromosomal analysis was normal and no recognized syndrome was felt to be present. She had an uncomplicated neonatal course. At age 14 months she was first noted to have a heart murmur and was found on investigation to have a double-outlet right ventricle with a subpulmonic ventricular septal defect and evidence of advanced pulmonary vascular obstructive disease. Her cardiac lesion was felt to be irreparable and no surgery was recommended. At age 25 months she was investigated for the onset of a new murmur and was found to have severe congenital mitral valve stenosis. During follow-up the mitral valve murmur increased in intensity and at age 11 years she underwent cardiac catheterization. The mitral stenosis was felt to be severe, with left atrial mean pressures of 27 mmHg, the atrial septum was intact and pulmonary hypertension was noted with a pulmonary arteriolar resistance of 7.5 Wood units. During follow-up she required no medications, although her level of physical activity decreased progressively. By age 20 years she had developed orthopnea, dyspnea on exertion, and headaches. One week prior to presentation she developed an exacerbation of her dyspnea and cough with increased cyanosis. She was admitted to a community hospital where her attending physician performed an echocardiogram. She was found to have a freely mobile mass in the left atrium. She was therefore transferred upon discharge to our institution for evaluation.

On arrival, she appeared to be alert, cyanotic, and dyspneic with marked fatigue. She had 2+ pitting edema of the lower extremities. There was a prominent right ventricular heave. On auscultation the first heart sound was loud, the second heart sound was loud and single, a gallop rhythm was noted with an intermittent mid-systolic click, and there was a grade I/VI soft ejection systolic murmur at the lower left sternal border with a grade III/VI mid-diastolic murmur radiating to the apex. An echocardiogram confirmed a normal-sized left atrium with some thickening of the wall, a round echogenic mass freely mobile in the cavity of the left atrium without apparent attachments, and severe congenital mitral valve stenosis with a small thickened poorly mobile mitral valve (Fig. 1). Her hemoglobin was elevated to 169 g/L with a hematocrit of 57%. She was admitted to the hospital and started on intravenous heparin therapy to prevent further clot formation. Streptokinase was not felt to be indicated given the probable age of the clot and the possibility of clot fragmentation with embolization. On the first day of admission she had a syncopal episode with a prolonged recovery of several hours, following which she was noted to have a visual field deficit with decreased acuity in the left eye and a mild right facial palsy. An ophthalmologic evaluation revealed a central retinal artery occlusion secondary to embolization. Magnetic resonance imag-

Address offprint requests to: Dr. Brian W. McCrindle, Variety Children's Heart Centre, FE241, Community Services Building, Winnipeg, Manitoba R3E 0Z2, Canada.

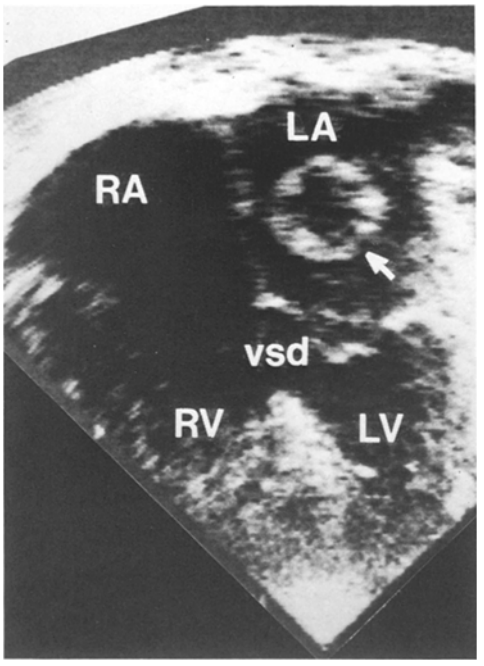

Fig. 1. Apical four-chamber two-dimensional echocardiographic view demonstrating the presence of a left atrial ball thrombus (*arrow*). *LA*, left atrium; *LV*, left ventricle; *RA*, right atrium; *RV*, right ventricle; *vsd*, ventricular septal defect.

ing of the head showed no abnormalities. She was therefore taken to the operating room urgently where she was found to have left atrial mean pressures between 35 and 40 mmHg, equal systemic and pulmonic arterial pressures, and a 2-cm ball thrombus free in the left atrium with a smaller 1-cm thrombus adherent to the septal wall. The thrombi were removed and the atrial septum excised to decompress the hypoplastic left atrium. The mitral valve appeared to be hypoplastic, thickened, and severely obstructed. Histology of the atrial septum showed moderate endocardial fibrosis. Postoperatively, she had an uneventful recovery with marked improvement in her fatigue and level of cyanosis. No anticoagulation therapy was given. A cardiac catheterization performed 2 weeks postoperatively showed continued severe pulmonary hypertension with advanced pulmonary vascular obstructive disease. The left and right atrial pressures were equal and low. She was discharged home much improved with possible heart–lung transplantation to be considered in the future.

Discussion

Intracardiac masses occur rarely in the pediatric population. A few reports have described the echocardiographic detection of a wide range of pathology, most frequently benign or malignant tumors arising either from intracardiac origin or from extension of intraabdominal tumors, or an intracavitary thrombus or vegetation [8]. Thrombi have been reported to occur under a variety of circumstances in each of the cardiac chambers. Some inciting prerequisites appear to be the presence of a thrombogenic surface, stasis of blood flow, and an abnormal hypercoagulable state. Right atrial thrombi have been reported in association with the presence of central venous catheters, intracavitary thromboembolism, the Fontan procedure, and the presence of vegetations due to endocarditis [2, 4, 6, 8]. Right ventricular thrombus has been found in one patient after a pulmonary valvotomy for pulmonary valve stenosis [8]. Left ventricular thrombi have been described in association with calcified ventricular fibromata, dilated cardiomyopathies, left ventricular fibroelastosis, and endomyocardial fibrosis [3, 8, 11].

Left atrial thrombi are more commonly described in adults with rheumatic mitral valve disease [7, 9]. However, there have been a few case reports of left atrial thrombus occurring in infants, all patients having associated mitral atresia and intact atrial septum [1, 5, 12]. Our patient had severe congenital mitral valve stenosis, a hypoplastic left atrium with an intact atrial septum, and mild polycythemia that may have further contributed to vascular stasis and thrombogenesis in the left atrium.

Increased vigilance on the part of pediatric cardiologists is necessary for the antemortem detection of this rare complication in this unique clinical setting. Early detection of left atrial thrombus is important before serious embolic phenomenon occur. Two-dimensional echocardiography remains the method of choice, with a reported specificity of 99% but a sensitivity of only 75% [9]. The use of transesophageal echocardiography may further improve detection of left atrial thrombi, particularly in the left atrial appendage. Medical management alone is inadequate, and left atrial thrombus has been reported to occur in an adult patient with mitral stenosis during adequate treatment with oral anticoagulants [10]. Embolic phenomena are very common if the left atrial thrombus is mobile [7]. Surgical removal of the thrombus and repair or palliation of the congenital heart disease is necessary in order to relieve the mechanical disturbance inciting thrombus formation and to prevent embolization. The presence of a left atrial ball thrombus in this setting must be considered a surgical emergency.

References

1. Beitzke A, Machler H, Stein JI (1987) Mitral atresia with premature closure of the oval foramen, right-sided levoatriocardinal vein and thrombus formation in the left atrium. *Int J Cardiol 14*:221–224

2. Felner JM, Churchwell AL, Murphy DA (1982) Right atrial thromboemboli: clinical, echocardiographic, and pathophysiological manifestations. *J Am Coll Cardiol 4*:1041–1051
3. Kupferschmidt C, Schmaltz AA, Tacke E, Apitz J, Lang D (1984) Left ventricular thrombi in three children with dilated cardiomyopathy: diagnostic procedure and clinical course. *Pediatr Cardiol 5*:65–70
4. Riggs T, Paul MH, DeLeon S, Ilbawi M (1981) Two dimensional echocardiography in evaluation of right atrial masses: five cases in pediatric patients. *Am J Cardiol 48*:961–966
5. Romano A, Weinberg PM, Woolf PK, Vetter VL (1989) Pulmonary venous obstruction from left atrial thrombus in hypoplastic left heart syndrome. *Pediatr Cardiol 10*:105–107
6. Saner HE, Ansinger RW, Daniel JA, Elsperger KJ (1984) Two-dimensional echocardiographic detection of right-sided cardiac intracavitary thromboembolus with pulmonary embolus. *J Am Coll Cardiol 4*:1294–1301
7. Schechter DC (1982) Left atrial ball-valve thrombus. *NY State J Med 82*:1831–1838
8. Sharratt GP, Lacson AG, Cornel G, Virmani S (1986) Echocardiography of intracardiac filling defects in infants and children. *Pediatr Cardiol 7*:189–194
9. Shrestha NK, Moreno FL, Narciso FV, Torres L, Calleja HB (1983) Two-dimensional echocardiographic diagnosis of left atrial thrombus in rheumatic heart disease. A clinicopathologic study. *Circulation 67*:341–347
10. van Dorp WT, vanden Berg BJ, van Rees C (1987) Left atrial ball thrombus during treatment with oral anticoagulants for more than one year. *Neth J Med 31*:16–19
11. Wiseman MN, Giles MS, Camm AJ (1986) Unusual echocardiographic appearance of intracardiac thrombi in a patient with endomyocardial fibrosis. *Br Heart J 56*:179–181
12. Wolf WJ (1986) Echocardiographic detection of a left atrial thrombus in an infant with complex congenital heart disease. *Am Heart J 112*:624–626