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Brief Report

Intra-atrial thrombus in a neonate with coarctation of the aorta

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Abstract It is uncommon for thrombus to form within the heart of neonates with congenital cardiac disease. We describe a newborn with coarctation of the aorta, in whom a left atrial thrombus was discovered on the second day of life, and was thought to have been present before birth.

Keywords: Thrombosis; hypoplastic aortic arch; congenital heart disease

Intra-cardiac thrombus is rare, and when encountered, is difficult to differentiate from other masses seen by echocardiography. The differential diagnosis includes primary and secondary cardiac tumours, which are extremely rare, and usually benign.[1] Thrombus should always be considered in the differential diagnosis. An intra-cardiac thrombus can appear cyst-like on echocardiography.[2] Although there are many predisposing conditions to its formation at this age, little is known about the exact aetiology, physiology, and natural history of the lesion as found in neonates.[3] Most of the evidence for its management is extrapolated from experience in adults.[4]

Case report

A female infant was born at term by lower segment caesarian section, weighing 3.9 kilograms. Her Apgar scores were 8 and 9 at one and five minutes, respectively. At two days of age, examination prior to discharge from hospital revealed that she had a systolic ejection murmur over her right upper sternal border, and she was referred to our cardiology service. Her physical examination showed a non-distressed and well-looking baby. Her vital signs were stable, with a heart rate of 130 beats per minute, respiratory rate of 50 per minute, blood pressure of 81 over 39 millimetres of mercury in the left arm, and a saturation of oxygen of 98 percent in room air. Her lungs were clear. The cardiovascular examination revealed a harsh systolic ejection murmur of grade 2 from 6. Her abdomen was soft, with the liver palpable 1 centimetre below the right costal margin. An echocardiogram showed preductal coarctation, with mild hypoplasia of the aortic isthmus, the arch opposite the origin of the left common carotid artery having a diameter of 3.4 millimetres, and a maximum Doppler velocity of 1.8 meters per second. The non-restrictive duct remained patent, permitting right-to-left shunting. The right ventricle and pulmonary trunk were dilated, with a regurgitant jet of 4.3 metres per second across the tricuspid valve. The right ventricular pressure was estimated at 85 millimetres of mercury. The left atrium was dilated, with aneurysmal dilation of the inter-atrial septum, and a mass measuring 1 by 1.7 centimetres was visualized in the left atrial appendage (Fig. 1). The mass appeared homogeneous, non-mobile, and confined to the appendage. There was a small patent oval foramen, permitting left-to-right shunting. Left ventricular function was normal.

The following day, the patient underwent a left thoracotomy, and the coarctation was repaired by means of an end-to-end anastamosis. The mass was monitored with serial echocardiography. Low molecular weight heparin was started on the third post-operative day. Three days later, a repeat echocardiogram showed that the mass had flattened, elongated, and was mobile, popping in and out of the mitral valve (Fig. 2). We

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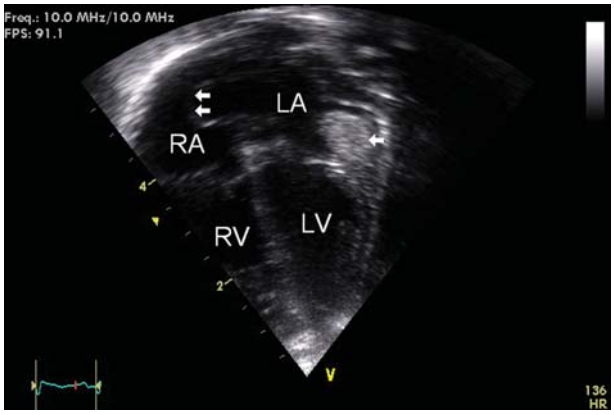

Figure 1.
Apical four-chamber view of an echocardiogram showing the left atrial thrombus (single arrow) and atrial septal aneurysm (double arrow). LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle.

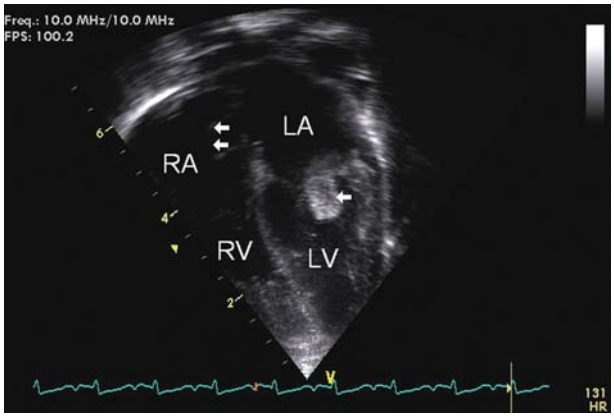

Figure 2.
Apical four-chamber view of an echocardiogram showing the left atrial mass (single arrow) extending through the mitral valve into the left ventricle. The atrial septal aneurysm is also visualized (double arrow). LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle.

were concerned that the mass would embolize and cause a cerebral accident, so the baby was taken to the operating room. Under cardiopulmonary bypass, the mass was removed through the left atrial appendage, the atrial appendage excised, and the atrial septal aneurysm resected. Histologic examination of the mass revealed an organized thrombus. The prothrombotic work-up, including assessment of protein C, protein S, antithrombin 3, Factor 5 Leiden, and the prothrombin gene, was within normal limits. Post-operatively, the patient developed pulmonary hypertension, which required treatment with nitric oxide and sildenafil for six days before resolving. Ten days later, she was sent home in stable condition on low dose aspirin, with good peripheral pulses and normal blood pressure. There was no evidence of residual or recurrent thrombus. At follow-up after three months, she was well and remained normotensive. Her echocardiogram revealed excellent repair of the coarctation, with no evidence of recurrent thrombus.

Discussion

The differential diagnosis of an intra-cardiac mass may include cardiac tumours, as well as thrombus. Before 12 months of age, the most common tumours are rhabdomyomas, teratomas, and fibromas, while atrial myxomas are extremely rare.[5] Of the primary malignant tumours, fibrosarcomas and rhabdomyosarcomas are the most common, with secondary malignant tumours, such as lymphosarcomas, being very uncommon.[5] Persistent blood-stained pericardial effusion is a common finding in the setting of malignant tumours.[5] The formation of intra-cardiac thrombus in infants and children, even those with congenital cardiac disease, is most uncommon in the absence of central catheters.[6] Newborns are at greatest risk, with the incidence of intra-cardiac thrombus being higher in those who are premature or born with low weight.[6] Conditions that predispose this group of patients to developing thrombus include maternal diabetes, perinatal asphyxia, sepsis, dehydration, autoimmune disease, the use of central lines, lower concentrations of antithrombin, heparin cofactor II, protein C, reduced fibrinolytic capacity, and high hematocrit.[4] The incidence drops significantly after the first year of life. In our case, we found no predisposing features, although aortic coarctation and aneurysmal dilation of the atrial septum were risk factors. There may have been stasis of blood in the left atrium due to reduced cardiac output, although contractility of the left heart appeared normal. One of the catastrophic complications of an intra-cardiac thrombus is systemic embolization, especially if it occurs intra-cranially.[3] This was avoided in our case by prompt surgical excision of the thrombus. We suggest that our case may be unique because the thrombus was most likely congenital, and was associated with aortic coarctation. To our knowledge, congenital left atrial thrombus has not previously been reported in this setting. Thrombus in the left atrial appendage is sometimes difficult to diagnose with echocardiography. In our case, nonetheless, echocardiography was useful in both detecting the thrombus and showing its size, location, and serial changes over time. Although atrial fibrillation and embolic events are known to be associated with congenital aneurysms of the atrial wall,[7] we doubt that the atrial septal aneurysm contributed to formation of the thrombus in our patient, since it was remote from the aneurysm, and the patient did not have any

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arrhythmias. The Canadian and International Registry of Neonatal Thrombosis classifies treatment of thrombus in four major categories: supportive care, anticoagulation, fibrinolysis, and surgery.[8,9] Hartmann et al.[6] successfully treated 11 out of 14 neonates with catheter-related thrombus with high dose recombinant tissue plasminogen. Herron et al.[10] reported two cases with acute left atrial thrombus treated successfully using low dose recombinant tissue plasminogen activator in combination with heparin, with no significant haemodynamic side effects.[10] We were concerned about the possibility of systemic embolization with thrombolytic therapy, and decided to remove the thrombus surgically. Although neonatal thrombotic disease has been treated with a wide range of agents, administered through various routes, and with varying dosage and duration of treatment, as yet there is no consensus about the most effective treatment.[6]

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