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Brief Report

Successful treatment of a thrombus in the left aortic coronary sinus in a child with systemic lupus erythematosus

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Abstract We report our experience in a 12 year old boy referred with suspected myocardial infarction. He has previously been diagnosed with systemic lupus erythematosus, and was being treated with steroids. Echocardiographic examination revealed a thrombus in the left aortic coronary sinus of Valsalva partially occluding the orifice of the left coronary artery. The thrombosis was successfully treated by venous thrombolysis using recombinant tissue plasminogen activator.

Keywords: Collagen diseases; interventional catheterisation; paediatrics

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Patients with systemic inflammatory diseases and antiphospholipid antibodies are prone to thrombotic complications.[1] Thrombosis of the ascending aorta, although extremely rare, is a possible cause for ischaemic cardiac injury. Most cases reported thus far have been adults with preexisting atherosclerotic disease, the majority being treated with open heart surgery.[2–4] We report our experience with a boy with acute myocardial ischaemia in whom we found a large thrombus obstructing the orifice of the left coronary artery. He was successfully treated with intravenous thrombolysis.

Case report

A 12-year old boy was transferred to our tertiary clinic for paediatric cardiology with a suspicion of an acute myocardial infarction. He had been diagnosed 20 days previously with systemic lupus erythematosus. He had been treated with corticosteroids, with good initial effect. After experiencing acute precordial pain, electrocardiographic signs of myocardial ischemia were noted, and he was transferred to our clinic.

On admission, the patient was in poor condition, with clinical signs of congestive cardiac failure. His heart rate was 120 beats per minute, he was breathing at a rate of 40 breaths per minute, exhibiting orthopnea, and his liver was enlarged. There were clinical and X-ray signs of pulmonary oedema. The electrocardiogram indicated acute myocardial ischemia, with elevated ST-segments in leads II, III, aVF, V5, and V6 (Fig. 1). Laboratory markers for myocardial cytolysis were positive. Transthoracic echocardiography revealed a small pericardial effusion and a structurally normal heart. The posterior left ventricular wall was hypokinetic and the global systolic left ventricular function was decreased, the ejection fraction being measured at 40%. An abnormal structure measuring 15 by 8 millimeters was noted in the ascending aorta just above the left coronary aortic sinus of Valsalva (Fig. 2). The aortic valve, nonetheless, functioned well, with no signs of stenosis or regurgitation. The structure was interpreted as a thrombus, directly adjacent to and probably partially obstructing the orifice of the left coronary artery, and deemed to be the reason for the myocardial ischaemia. Intravenous thrombolysis with recombinant tissue plasminogen activator was initiated. We initially gave a bolus of 0.5 mg/kg Actilyse®, and followed this with a perfusion of 1 mg per kg over 24 hours. Transthoracic echocardiography repeated 3 hours after administration of the bolus showed complete resolution of

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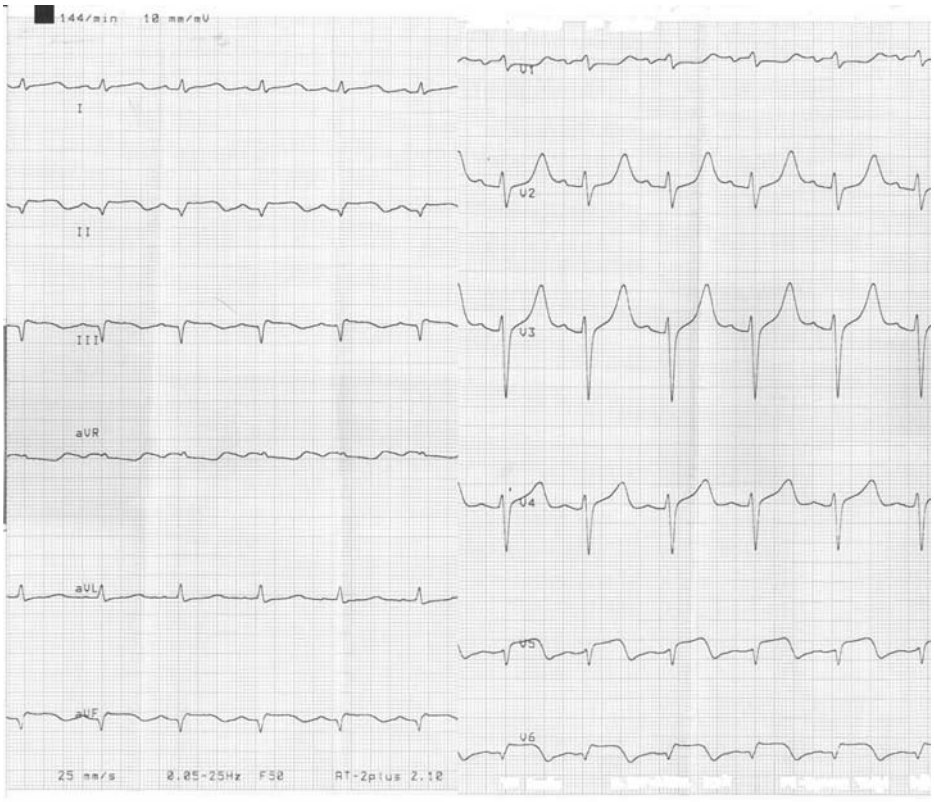

Figure 1.
The electrocardiogram shows ST-elevations indicative of ischaemic changes.

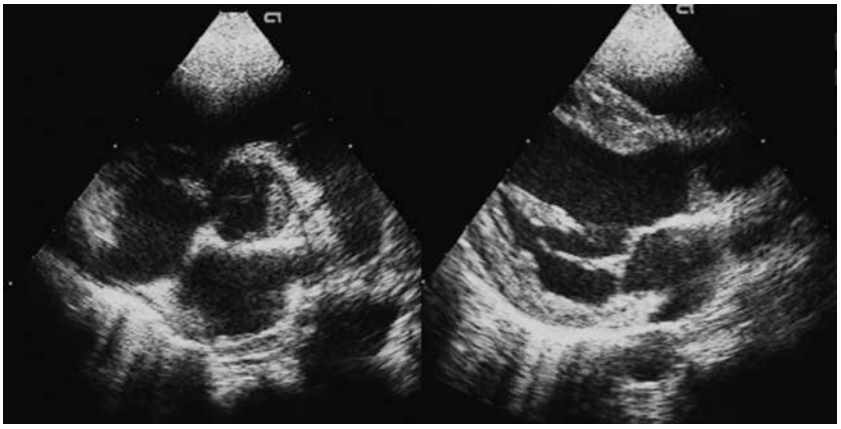

Figure 2.
The echocardiograms in short and tilted long axis views show a thrombus in the left aortic sinus of Valsalva opposite to the orifice of the left coronary artery.

the thrombus. No bleeding or embolic events associated to the thrombolytic therapy were detected. The thrombolysis was discontinued, and heparinisation continued, the target being a ratio of the activated partial thromoplastin time between 2 and 2.5. Over the next 3 days, his condition improved dramatically. The congestive cardiac failure was reduced, and the levels of the biomarkers in the serum normalised. Transthoracic echocardiography showed no thrombotic or other abnormal structures in the heart and great vessels. He was discharged on oral anticoagulation therapy. At follow up after 1 month, there were no clinical signs of congestive cardiac failure. Echocardiography showed no abnormal structures in the heart and great vessels. The global systolic function, nonetheless, remained reduced, and the posterior left ventricular wall was still hypokinetic.

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Discussion

It is well known that patients with autoimmune diseases, particularly systemic lupus erythematosus, are at an increased risk of thrombosis throughout the course of the disease. In a recent follow up of adults with this disease, the incidence of thrombosis was 36.3 per 1000 patient-years.[1] Thrombosis within the aortic sinuses of Valsalva, however, is an extremely rare condition. It has been reported mainly in isolated adults, resulting in acute myocardial infarction because of partial obstruction of the orifices of the left or right coronary arteries.[2–4] Ours is, to the best of our knowledge, the first report of thrombosis within an aortic sinus of Valsalva causing myocardial ischaemia in a child with systemic lupus erythematosus. The presence of a thrombotic formation is extremely unexpected in the ascending aorta in the setting of normal structure and flow. In most previously reported cases, such thromboses were thought to be associated with an atheromatous plaque.[4] Such an explanation is unlikely in our young patient. Although being predisposed for thromboses in the setting of the antiphospholipid antibodies known to be associated with systemic lupus erythematosus[5], we still believe that there has to be some abnormality in the endothelium of the aortic wall. Possible underlying conditions could be Libman-Sacks endocarditis with small vegetations not detectable by transthoracic echocardiography, or inflammatory changes in the aortic wall due to aortitis, as described by others.[6]

Most patients with ascending aortic thrombosis have been treated with open heart surgery.[4,6] Another theoretical possibility for treatment could be conservative anticoagulation followed by eventual spontaneous thrombolysis. To our knowledge, there is only one previous case reported with successful intravenous thrombolysis of a thrombus of the ascending aorta.[2] Even this approach was thought to be hazardous because of possible major systemic embolic complications.[3] Our patient was treated conservatively because of the possible risks associated with emergency cardiac surgery, and the increasingly good reported results of thrombolysis in children.[7] We also could not wait for spontaneous thrombolysis, as the patient was in a critical condition. In retrospect, our decision seems to be justified. The thrombus was resolved without any detectable side effects, and the surgical trauma was avoided.

In most previous reports, the thrombus within the sinuses of Valsalva has been diagnosed by transoesophageal echocardiography or angiography.[2,4] We relied only on transthoracic echocardiography, thus avoiding the additional risks of these methods. Our patient, however, was young and thin, permitting us to make good images. In summary, we report a rare case of a thrombosis in the sinus of Valsalva causing myocardial ischaemia in a child with systemic lupus erythematosus. He was treated successfully using recombinant tissue plasminogen activator without any side effects.

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