Giant right atrial diverticulum: an unusual cause of Wolff-Parkinson-White syndrome

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Abstract
A case of a giant right atrial diverticulum associated with neonatal supraventricular tachycardia is reported. The electrocardiogram in sinus rhythm showed pre-excitation that may have been caused by the right atrial diverticulum adhering to the right ventricle.

Congenital diverticulum or aneurysm of the right atrium is a very rare anomaly. Morrow et al reported the case of a 23 year old woman with a large congenital aneurysm of the right atrium that caused repeated attacks of supraventricular tachycardia and was successfully treated by surgical resection of the diverticulum. A case of atrial tachycardia in a five month old infant caused by multiple congenital sacular aneurysms of the right and left atrium that was successfully treated surgically has also been reported. Aneurysms of the left atrium have also been reported to be associated with atrial arrhythmias. Our case was noticed prenatally, was subsequently diagnosed with cross sectional echocardiography, and was successfully treated with antiarrhythmic drugs.

Case report
A 29 year old woman (para 1, gravida 3) was referred at 35 weeks’ gestation after a four chamber view of the fetal heart could not be obtained. Ultrasound examination of the heart showed an unusually dilated right atrium and right ventricle with normal attachment of the septal tricuspid valve leaflet. No other cardiac abnormality was found to explain the right atrial and ventricular enlargement.

Karyotyping of a sample of fetal cord blood showed normal chromosomes. In view of cardiac abnormality detected, delivery was induced at 38 weeks’ gestation at the regional neonatal unit. A male baby weighing 3 kg was delivered normally and was in good condition at birth. Within two hours of birth he developed supraventricular tachycardia at a rate of 275 beats per minute. He was treated with oral digoxin and intravenous flecainide and the heart rate slowed to 225 beats per minute (fig 1A). The following day because the arrhythmia had persisted he was given 5 J and then 10 J of direct current synchronised cardioversion without effect. The next day he converted to sinus rhythm with a short PR interval and a delta wave with a pattern typical of type B Wolff-Parkinson-White syndrome (fig 1B).

Echocardiography, performed when supraventricular tachycardia first developed, showed a large diverticulum (fig 2) arising from the lateral right atrial wall and extending over the right ventricle to its apex. The wall of the diverticulum seemed to be trabeculated internally and to contract during the tachycardia. Strands of muscle tissue crossed the diverticulum. The right ventricular free wall was concave and it looked as if the right ventricle was being compressed by the diverticulum. However, the tricuspid valve measurements were within normal limits and there was no disturbance of Doppler flow patterns within the right ventricle. Contrast echocardiography performed through a peripheral venous line showed microbubbles entering the diverticulum where they seemed to persist and whirl about. A chest x ray showed a very prominent right atrial shadow. He was discharged home on flecainide and digoxin. During follow up for one year he thrived and had no recurrence of supraventricular tachycardia. His electrocardiogram continues to show type B Wolff-Parkinson-White syndrome. Repeated echocardiography and ventilation/perfusion scans showed no evidence of thrombus formation within the diverticulum or embolism to the lungs.

Discussion
Neonatal supraventricular tachycardia is generally associated with a structurally normal heart and is usually caused by an accessory pathway. When neonatal supraventricular tachycardia is associated with congenital heart disease the most likely diagnosis is Ebstein’s anomaly, which can be associated with type B Wolff-Parkinson-White syndrome. In our patient enlargement of the right atrium caused by a diverticulum was congenital. There was no evidence that this diverticulum was secondary to tricuspid valve regurgitation. On echocardiography the aneurysm seemed to arise from the free wall of the right atrium and extended across the front of the heart to reach the left ventricular apex. No distinct right atrial appendage was seen. The wall of the diverticulum seemed to be muscular and contracted with atrial systole. The patient of Morrow and Behrendt had a structurally similar aneurysm in terms of origin and disposition on the cardiac surface. Histology of the resected aneurysm in their patient showed that the wall of the
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Figure 2. Subcostal four chamber cross-sectional echocardiogram showing right atrial diverticulum in front of the compressed right ventricle (arrow). RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle.

Figure 1. (A) Electrocardiogram showing narrow QRS complex tachycardia at a rate of 225 bpm, with P wave visible in the ST segment suggesting atrioventricular reentry. (B) Electrocardiogram in sinus rhythm showing a short PR interval and delta wave, characteristic of type B Wolff-Parkinson-White syndrome.

diverticulum was composed of fibrous tissue and attenuated muscle fibres with an endothelial lining. The atrial diverticulum in their patient case was easily reflected off the ventricles suggesting that there was an intervening serous space lined by visceral pericardium. The echocardiogram in our patient showed that the diverticulum was directly adherent to the anterior surface of the right ventricle and left ventricle and that there seemed to be large muscle bundles arising from the ventricular aspect of the diverticulum that protruded into the cavity of the diverticulum. The right ven-

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