Echocardiographic diagnosis of multiple congenital aneurysms of the sinus of Valsalva

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SUMMARY Aortic regurgitation developed in a 56 year old man with severe nephrotic syndrome. Cross sectional echocardiography showed bilateral aneurysms of the sinus of Valsalva and a bicuspid aortic valve. He died of intercurrent pneumonia. Postmortem examination confirmed the presence of the congenital aneurysms.

Congenital aneurysm of the sinus of Valsalva usually affects only one aortic sinus; however, cases of multiple congenital aneurysms of the sinus of Valsalva have been reported.¹ Cases of acquired aneurysm are seen in endocarditis and syphilis. Several reports have suggested that aneurysms of the sinus of Valsalva can be diagnosed by echocardiography.^{2,3}

We report a case of congenital bilateral aneurysms of the sinus of Valsalva in a patient with a bicuspid aortic valve.

Case report

A 56 year old man presented with a one week history of ankle and facial swelling. He had a short history of anorexia and had lost a few pounds in weight.

On examination he looked well. His blood pressure was 110/60 mm Hg and he had a grade 2/6 early diastolic murmur in the aortic area. The jugular venous pressure was not raised. He had pitting oedema of the ankles and dipstick testing of his urine showed that it contained blood and a large amount of protein.

The diagnosis of an acute nephritis associated with the nephrotic syndrome was confirmed (serum urea 29.9 mmol/l and creatinine 260 μ mol/l and urinary protein excretion 58 g/l). Immunological studies including immunoglobulin and complement concentrations and antinuclear factor antibodies were normal. A renal biopsy specimen showed changes of focal segmental glomerulosclerosis.

Repeated sets of blood cultures were negative for bacteria and fungi. The electrocardiogram showed

left ventricular hypertrophy and his chest x ray showed moderate cardiomegaly.

Cross sectional echocardiography was performed with a Hewlett Packard 77020A real time phased array scanner. It showed a bicuspid aortic valve that was thickened and calcified. The excursion of the aortic valve was impaired. The parasternal sagittal view (figure a) showed bilateral aneurysms of the sinus of Valsalva. The first aneurysm, which was 2 cm in diameter, arose from the right coronary sinus and protruded into the right ventricular outflow tract. The second aneurysm arose from the left coronary sinus and protruded into the left atrium. The left ventricle was moderately hypertrophied and there was a small pericardial effusion.

Surgical correction of these aneurysms was prevented by the renal failure and massive protein loss (up to 80 g/day). After therapeutic renal artery embolisation he was maintained on regular haemodialysis, but a pneumococcal pneumonia developed and he died.

At necropsy the left ventricular hypertrophy and the presence of a bicuspid aortic valve were confirmed. The valve cusps showed fibrocalcific thickening and mild stenosis. There were bilateral aneurysms of the sinus of Valsalva (figure b). Both had a fibrous wall, contained organising blood clot, and communicated with the sinus via a defect lying between the cusp insertion and the coronary artery ostium. The right aneurysm of the sinus of Valsalva measured 2.5 $\times$ 1.5 $\times$ 3.0 cm and lay between the aortic root and the pulmonary infundibulum, with extension into the upper intraventricular septum. There was a rupture, 1.8 cm in diameter, through to the subvalvar area. The left aneurysm of the sinus of Valsalva measured 2.0 $\times$ 1.0 $\times$ 2.8 cm and lay between the aortic root, left atrium, and left atrial

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Figure Parasternal long axis echocardiographic view (a) and section (original magnification $\times 1.2$) (b) through the left ventricular outflow tract showing two aneurysms (An) of the sinus of Valsalva with rupture (Rup) of the right aneurysm into the left ventricle (LV). EP, entry points of aneurysms; A, aorta; RV, right ventricle; LA, left atrium; MV, mitral valve.

appendage. Rupture through to the subvalvar area was incipient.

Discussion

Aneurysms of the sinus of Valsalva are rare. To date there have been 276 cases reported in English.⁴⁻¹² Such aneurysms often present after rupture in adult life. The diagnosis may be suspected from a history of abrupt onset of symptoms including dyspnoea, chest pain, or palpitation in the presence of a continuous murmur.¹³ The mean age of presentation is 31 years. Most patients present with congestive cardiac failure and about 18% are symptom free at the time of detection. Aneurysms of the right sinus are frequently associated with a ventricular septal defect. The clinical manifestations of a ruptured aneurysm of the sinus of Valsalva depend on the direction and amount of blood flow through the abnormal intracardiac communication. The lesions are most commonly found in the right sinus and non-coronary sinus.⁴ Perforation into the right sided cavities is more common; however, rupture may also occur into the left side of the heart, the pulmonary artery, the superior vena cava, the pericardium, and the intraventricular septum.^{5,6}

In practice the diagnosis is usually confirmed at angiography; but several reports have shown that M mode and cross sectional echocardiography are valuable for non-invasive diagnosis.⁶⁻⁸ Rupture of an aneurysm into either the right ventricular outflow

tract or to the right atrium has been seen in parasternal views.⁶ In our patient the aortic sinuses from which the aneurysms originated could be identified on parasternal views, and a possible rupture into the left ventricle was noted. Unfortunately, Doppler echocardiography, which would have been of great value in confirming the presence of a communication between the aneurysm and the left ventricle, was not available to us at the time of the examination.

Aneurysms of the sinus of Valsalva arising from bicuspid aortic valves are very rare—only five cases have been reported,^{4,9,10} and rupture of a right sided aneurysm of the sinus of Valsalva into the left ventricle has been reported in only three cases.¹² We believe that ours may be the first report to describe the echocardiographic features of multiple congenital aneurysms of the sinus of Valsalva.

We thank Dr D H Bennett for his constructive criticism during the preparation of this paper.

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