

Aneurysm of the coronary sinus

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ABSTRACT Two patients, an adult and a baby, with an aneurysm of the coronary sinus are described. This unusual anomaly was detected in the baby during angiographic investigations for congenital heart disease. The aneurysm was an unexpected postmortem finding in the adult. Although the adult died suddenly, there was no evidence that this could be linked to the presence of the aneurysm.

An unusual malformation of the heart forms the basis of this report. We have recently encountered two patients in whom a pouch from the floor of the coronary sinus extended subendocardially into the diaphragmatic surface of the ventricular mass. Although superficially giving the impression of a supernumerary ventricle, the anomaly is an atrial aneurysm. We are unaware of any prior description of such a lesion.

Case histories

CASE 1

A baby boy, born at term after a normal pregnancy and caesarean section, was first seen at the age of 1 year. His weight and height were well below the 10th percentile. At that time he was somewhat tachypnoeic and was wheezing. A month later he was admitted to hospital for further investigations. He was pink at rest with no evidence of heart failure. There was a normally split second sound with a loud pulmonary component and a grade 3 ejection systolic murmur best heard in the second left intercostal space with no diastolic murmur. The chest radiograph showed appreciable cardiac enlargement and considerably increased pulmonary vascular markings. The electrocardiogram (ECG) showed sinus rhythm, a mean QRS frontal axis $+180^\circ$, and severe right ventricular hypertrophy. Cross-sectional echocardiography showed normally arranged chambers with atrioventricular concordance and ventriculoarterial concordance. A single

large perimembranous trabecular ventricular septal defect was detected at angiography. An injection into a persistent left sided superior caval vein showed contrast streaming into the coronary sinus and the right atrium. It also showed an aneurysm extending from the coronary sinus into the diaphragmatic aspect of the ventricles (fig 1). This aneurysm was smaller in ventricular systole than in ventricular diastole. In view of the severe right ventricular hypertrophy and the non-pansystolic murmur we believed there to be a risk that this infant would develop pulmonary vascular disease if the ventricular septal defect was not repaired. The defect was surgically closed through the right atrium three months later. The postoperative course was uneventful. Eight weeks after the operation the patient looked well and was thriving and symptom free.

CASE 2

A 23 year old man, a fitter by profession, was performing routine exercises at a karate club when he suddenly looked pale and fell to the floor. Resuscitation attempts were unsuccessful and he was certified "dead on arrival" in the hospital casualty department. His past history was unremarkable, apart from one attack of fainting seven years earlier. On that occasion he had also been training for karate and later complained of discomfort in the chest and palpitations. He felt cold and sweaty. He was admitted to hospital, where an ECG showed signs of tachycardia and inverted T-waves in leads III and AVF. A diagnosis of tachycardia of unknown aetiology was made. The arrhythmia settled after administration of lignocaine. The chest radiograph and serum enzymes were normal. He was kept

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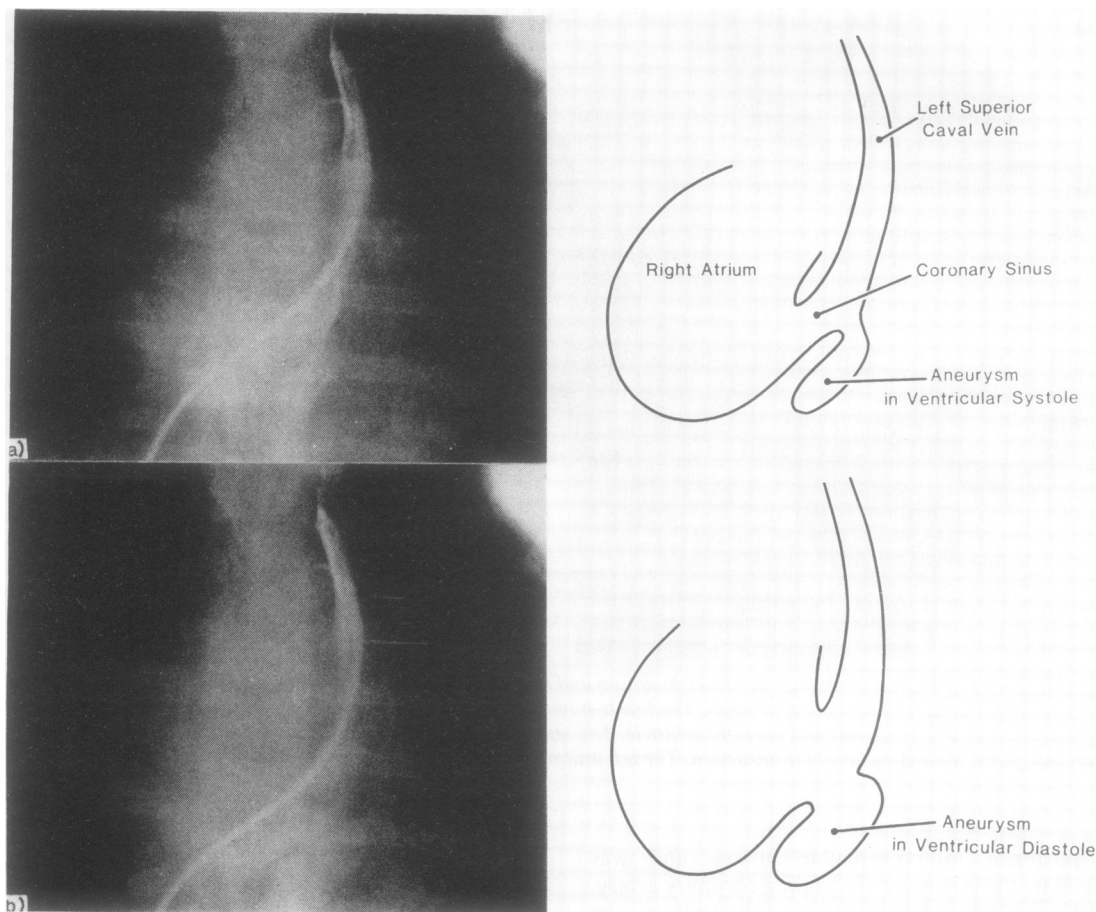


Fig 1 Two frames taken during angiography showing an injection of contrast into a persistent left sided superior caval vein. The line diagrams were traced from angiograms (a) and (b) to show the aneurysm as seen in ventricular systole and diastole respectively.

under hospital observation for a few days, during which the ECG showed episodes of bradycardia. There was no follow-up after discharge and the patient did not present with symptoms again until the fatal event.

At necropsy the heart weighed 300 g after fixation in formalin. The gross appearance of the heart was unremarkable apart from the position of the posterior descending coronary artery, which was more to the right and nearer to the marginal than usual. When the right atrium was opened a very unusual anomaly was seen. The atrial septum was intact, but the right atrium appeared to drain to the ventricular mass via two orifices. One was guarded by the tricuspid valve and opened to the right ventricle. The second orifice was unguarded. In addition, the left atrium was connected normally to the left ventricle, this connection being guarded by the mitral

valve. Examination of the anomalous chamber showed that it originated from the floor of the coronary sinus and extended into the diaphragmatic aspect of the ventricular mass at its crux (fig 2a). The atrial opening was triangular in shape when viewed from above and the pouch extended about 6 cm towards the cardiac apex. On its endocardial surface the pouch showed pits and faint trabeculations in places. The left wall of the pouch was muscular, being continuous with the atrial septum superiorly (fig 2b). The atrial septum was deviated to the left posteriorly. The upper part of the chamber bulged into the left ventricle, producing a muscle ledge just beneath the annulus of the mural mitral valve leaflet. The inferior or parietal wall was very thin and appeared deficient in myocardium. On the right the wall was again muscular. The wall separating the anomalous chamber from the tricuspid valve ran to

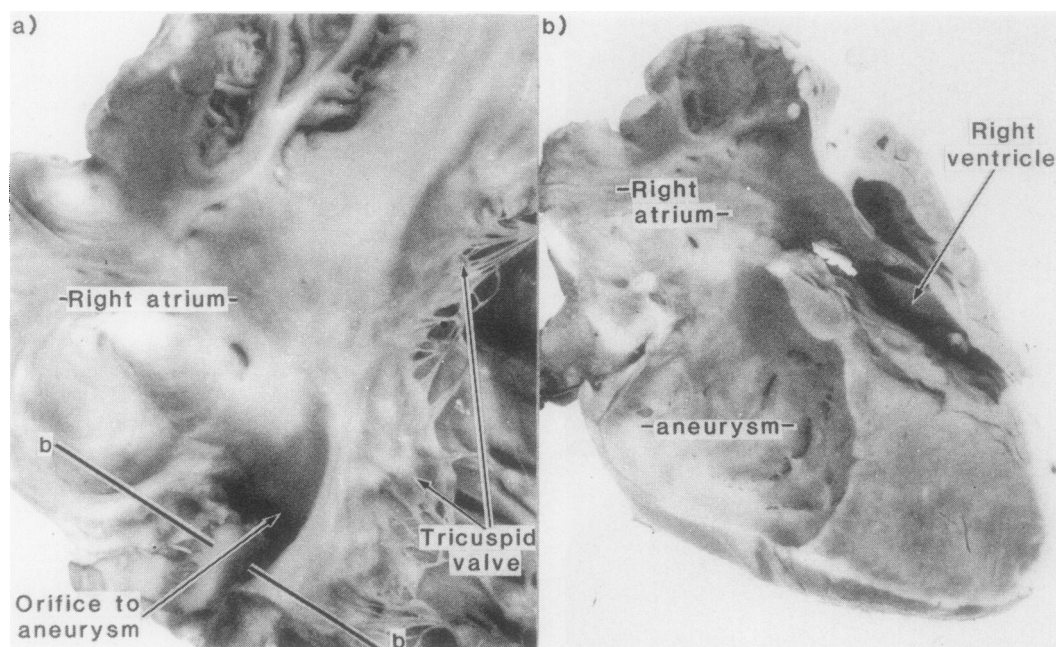


Fig 2 Two views of the heart from case 2. The right atrium and right ventricle have been opened in (a) to show the orifice of the aneurysm in the floor of the coronary sinus. The line b-b marks the plane of section shown in (b). The right atrium communicates with the aneurysm and the right ventricle. The septal leaflet of the tricuspid valve is attached to a muscular septum that forms the right wall of the aneurysm. The left wall of the aneurysm is continuous with the atrial septum.

the posterior interventricular furrow and was marked by the posterior descending coronary artery. Apart from the extension into the walls of the pouch, the right and left ventricular chambers were normally formed, although the right chamber was somewhat smaller than anticipated. The ventricular outlets were normal and supported their appropriate great arteries.

Histological sections were made of the parietal wall and the right wall of the anomalous chamber. The parietal wall contained only a thin strip of atrial myocardium sandwiched between some fibrous and fatty tissue and the endocardium and epicardium on either surface (fig 3a). The basal part of the right wall was considerably thicker and the section showed a rim of myocardium on the atrial aspect separated from ventricular myocardium by the fibrous tissue of the tricuspid valve annulus (fig 3b).

Discussion

To the best of our knowledge, the anomaly present in these hearts has never been reported before. Although at first sight presenting as a supernumerary ventricular chamber, the pouch is in reality an

aneurysm of the coronary sinus. This is shown by its having a wall composed of atrial musculature, the entire pouch being on the atrial aspect of the atrioventricular junction despite its location within the diaphragmatic surface of the ventricular mass. As expected from this anatomy, the muscular tissue between pouch and right ventricle is part of the inlet ventricular septum. In keeping with this, the posterior descending coronary artery runs down its epicardial aspect. In the first of our cases the observation of the aneurysm was a chance finding at angiography. It attracted our attention because we were in the process of studying the fatal case. We do not anticipate the aneurysm to be of clinical significance, although the sudden death did give cause for alarm as the pouch was discovered unexpectedly at necropsy. The extension into the diaphragmatic surface had not disturbed the position of the atrioventricular conduction tissues, which were histologically normal. There is no good evidence therefore to link the aneurysm with the sudden death, but the presence of such a large supernumerary atrial compartment is striking.

On reflection, in view of the syncopal attack and cardiac arrhythmia after vigorous exercise in the

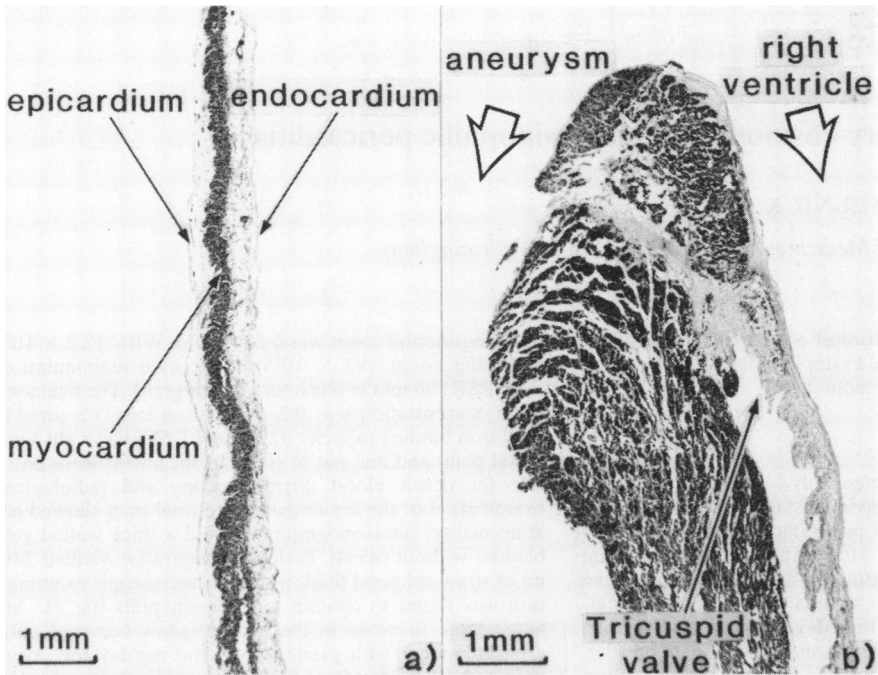


Fig 3 Photomicrographs of sections from the aneurysm in case 2: (a) a section from the parietal wall, shown in contrast to (b) a section from the right wall.

second patient, perhaps intracardiac electrophysiological investigations should have been carried out when the patient was first seen. This would have elucidated further the conduction abnormalities. The value of the information that may be obtained by this invasive procedure must, however, be carefully evaluated beforehand. Cardiac cineangiography performed at that time would certainly

have showed the aneurysmal pouch, although it is questionable whether this revelation on its own would have altered the course of management in this particular patient.

We thank our clinical colleagues who have looked after these patients.