Giant aneurysm of membranous septum

Unusual cause of mediastinal mass

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SUMMARY A case of a giant aneurysm of the membranous interventricular septum which presented as a mediastinal mass is described. Aneurysms of the membranous septum are rare and usually found incidental to the main cause of the patient’s complaints. They are most commonly associated with ventricular septal defects but can also be found in the presence of ventriculoarterial discordance, atrioventricular canal defects, and semilunar valve abnormalities. In patients presenting with a mediastinal mass which may be cardiac, full left and right heart catheterisation are necessary; a simple right heart study is inadequate.

Aneurysms of the membranous septum are occasionally associated with ventricular septal defects and other congenital heart lesions. A patient is described with a massive aneurysm of the membranous septum which presented as a mediastinal mass and which was successfully resected surgically.

Case report

A 36-year-old Pakistani civil servant was admitted for investigation of breathlessness and a mediastinal mass. Eight years before admission he was treated for infective endocarditis after recurrent fever. Six years before admission he was treated for two years with antituberculous drugs for histologically proven tuberculosis of the caecum. For the 18 months before admission he had become progressively breathless on exertion such that he had to rest after three flights of stairs. Five months before admission a chest x-ray film was found to be abnormal, having been unequivocally normal five years later.

On examination he looked well. In the cardiovascular system, the pulse was 80 per minute, regular, large in volume; the venous pressure was raised to 5 cm, the blood pressure was 140/70 mmHg. The heart sounds were normal, with a grade 3/6 ejection type systolic murmur best heard at the base but radiating into the back and a grade 3/6 early diastolic murmur best heard at the lower left sternal edge. The liver was not enlarged, there was no peripheral oedema, and the lungs were clear. Apart from a laparotomy scar, general examination was unremarkable. The electrocardiogram showed sinus rhythm 80 per minute and was within normal limits. Chest x-ray film (Fig. 1) showed a localised 6 cm round bulge on the right border of the heart, which on lateral and oblique

Fig. 1 Plain chest x-ray film showing localised bulge related to right heart border.
views was shown to lie on the anterolateral aspect of the right side of the heart. Echocardiography was attempted but was unsuccessful for technical reasons.

An initial diagnosis of mixed aortic valve disease with, in addition, a mediastinal mass was made. At fluoroscopy the mass was seen to pulsate but this was thought to be transmitted pulsation from the right atrium. A diagnosis of a pericardial cyst was made and a thoracotomy performed.

At the limited anterior right thoracotomy the mass was found to be covered by pericardium, to expand in time with ventricular systole, and to contain systolic and diastolic thrills. The ascending aorta was felt arising above the mass and medial to it and it was felt possible that the mass was an aneurysm of the sinus of Valsalva. In view of these findings no further surgery was attempted on this occasion and the chest was closed.

When the patient had convalesced from operation cardiac catheterisation was performed. Right atrial injection of contrast disclosed a large filling defect in this chamber and on the follow-through phase this was found to be in communication with the left ventricle. A separate left ventricular injection showed it to be an enormous aneurysm of the membranous septum (Fig. 2).

At a second operation the aneurysm was found to be pushing the right atrial appendage downwards and to be densely adherent to the pleura, the superior vena cava, and the junction of the superior vena cava and right atrium. When full extracorporeal circulation had been established the heart was electively defibrillated and a left ventricular vent was inserted. Suction of this vent collapsed the aneurysm. The aneurysm was opened and was seen to be trabeculated and to contain recent and old thrombus in its lateral pockets. The opening from the upper part of the septum immediately below the aortic valve was seen to measure about 7 mm in diameter with tough fibrous edges. This opening was closed with horizontal mattress sutures backed with Teflon and the aneurysm resected. Mild aortic regurgitation was present but the aortic valve was left alone.

After operation the patient made a good recovery and his chest x-ray film returned to normal, though a soft grade 1/6 early diastolic murmur remained audible at the left sternal edge.

Discussion

Aneurysms of the membranous septum are rare in postmortem series, occurring in four cases in 3000 necropsies reviewed by Rae1 and two cases in 16 000 necropsies reviewed by Steinberg.2 In Abbott’s account of 1000 cases of congenital heart disease,3 16 cases were described, in nine of which it was as an associated anomaly. Vidne et al.4 suggest that its incidence has been underestimated and state that it accounted for “10% of associated cardiac anomalies treated surgically”.

When present, aneurysms of the membranous septum are frequently associated with other cardiac defects, notably ventricular septal defects but also with transposition of the great arteries,4 corrected transposition,5 atroventricular canal defects,6 and abnormalities of the pulmonary and aortic valves, particularly aortic regurgitation.7 They are, often regarded as forming before the natural closure of ventricular septal defects8 and are usually found incidental to the main cause of the patient’s investigation. Occasionally, however, they are themselves the cause of important cardiac problems. The aneurysms usually pouch into the right side of the heart and may cause right ventricular outflow tract obstruction,9 10 particularly in the presence of transposition of the great arteries,4 5 and more rarely left ventricular outflow tract obstruction.11 Rupture of the aneurysm into the right heart giving a large left to-right shunt has been described,12 as has rupture into the pericardium with subsequent haemopericardium and death.13 They may be the site of infective endocarditis15 14 as was likely, though unproven, in our case, and as in our case they may be the site of thrombus formation.13 Cardiac arrhythmias and conduction defects of all varieties have been attributed to aneurysms of the membranous septum7 and two cases of complete heart block have also been re-
corded, one of which was treated effectively by permanent pacing, but the other died.16

Our case is similar to that of Saab et al.17 Their case also presented with ejection systolic and early diastolic murmurs and an abnormal shadow along the right cardiac border. In addition both their and our patient underwent exploratory thoracotomy before the correct diagnosis was established. These cases indicate the importance of complete right and left heart studies in cases of unusual anterior mediastinal masses of possible cardiac origin.

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References


2 Steinberg I. Diagnosis of congenital aneurysm of the ventricular septum during life. Br Heart J 1957; 19: 8–12.


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