Aneurysmal dilatation of left atrial appendage diagnosed by cross sectional echocardiography and surgically removed

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SUMMARY An isolated aneurysmal dilatation of the left atrial appendage was found in an 18 year old girl who presented with atrial fibrillation and an unusual cardiac shadow on routine chest radiographs. The diagnosis was made by cross sectional echocardiography. The giant appendage was excised to remove the risk of systemic embolism and the need for life long anticoagulation.

Case history

An 18 year old girl sought medical advice for headaches and was found to have an irregular pulse. Physical examination was otherwise normal. A chest radiograph showed a normal sized heart with a prominent left heart border (Fig. 1a), which on fluroscopy did not pulsate. The electrocardiogram confirmed atrial fibrillation. Treatment with anticoagulants was started. At 14 years old she developed the Guillain-Barré syndrome, and when in hospital a routine electrocardiogram showed sinus rhythm but no chest radiograph was taken.

Cross sectional echocardiography in the parasternal long axis view showed a normal sized left atrium and mitral valve. There was a narrow, long echo free space behind and lateral to the left ventricle. The space appeared to be continuous with the left atrium. In the apical four chamber view it was obvious that this "pouch" connected to the left atrium, extending along the free wall of the left ventricle (Fig. 1b). Computed axial tomography of the heart confirmed these findings and showed that the pericardium was intact over the aneurysmal atrial appendage. Cardiac catheterisation showed normal intracardiac pressures and no evidence of mitral valve disease. Pulmonary angiography indicated opacification of the shadow in the venous phase and contrast swirled in the "aneurysm" for 20 seconds. There was no visible filling defect in the appendage to suggest a mural thrombus (Figs. 2a and 2b). Amputation of the appendage was

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recommended to obviate the need for long term anticoagulants to prevent systemic emboli.

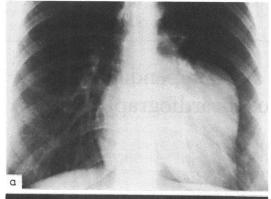
Through a left thoracotomy an enormous left atrial appendage was seen to compress and indent the left ventricle. The mitral valve was normal to palpation. The appendage was amputated at its base. Histological examination of the base of the left atrial appendage showed hypertrophied myocardium with an increase in interstitial fibrous tissue and thickening of the endocardium. Two weeks after operation the rhythm reverted spontaneously to sinus rhythm which has remained for 6 months without medication.

Discussion

Aneurysmal dilatation of the left atrium involving the appendage is usually associated with mitral valve disease. Herniation of the left atrial appendage through a pericardial defect may cause it to distend. In the absence of a known predisposing cause the condition has been assumed to be of congenital origin. Only one third of the patients, however, have presented symptomatically before the age of 10 years. Usually the problem is discovered in the second to fourth decades of life. Its discovery in a 10 month old baby³ suggests that it is of congenital origin or is acquired from a congenital lesion of the left atrial wall.

Histology of the appendage in other patients has shown a similar appearance to that in this case. Whether this results from damage to the atrial wall in intrauterine or postnatal life is not known.

The diagnosis is suggested by the appearance of the cardiac shadow on the chest radiograph. Other conditions which produce a similar shadow are mediastinal



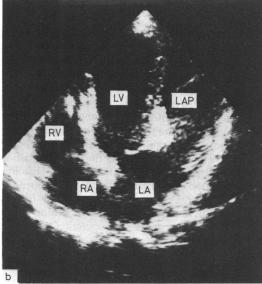


Fig. 1 (a) Chest radiograph in posteroanterior projection showing a large, rounded mass at the left heart border corresponding to the aneurysm of the left atrial appendage. (b) Cross sectional echocardiogram in the apical four chamber view showing an aneurysmal left atrial appendage (LAP). LV, left ventricle; RA, right atrium; RV, right ventricle.

and cardiac tumours or pericardial cysts. Angiography has been used to distinguish the condition. In our patient, cross sectional echocardiography but not M mode echocardiography clearly demonstrated the cause of the protuberance of the left heart border showing that the echo free space communicated with the left atrium, which differentiated it from a tumour and a pericardial cyst. Computed axial tomography of the heart was valuable in that it established the relation between the left atrial appendage and pericardium. Cardiac catheterisation was performed to exclude coexisting heart disease but was not necessary to establish the diagnosis.

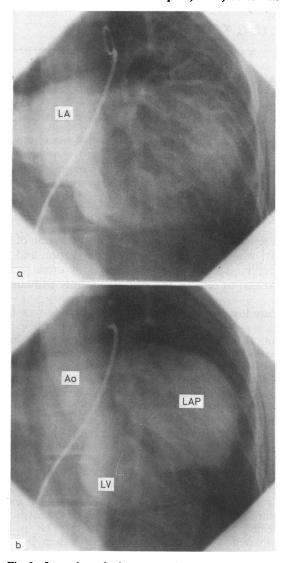


Fig. 2 Laevophase of pulmonary arteriogram: (a) contrast seen filling pulmonary veins and body of left atrium (LA) with appendage not opacified and (b) later frame with contrast filling left ventricle (LV) and aorta (Ao) but contrast still present in left atrial appendage (LAP).

Recognition of an isolated aneurysm of the left atrial appendage is important since supraventricular arrhythmias and systemic embolisation are common. The aetiology of the supraventricular arrhythmias is not understood. Perhaps they result from the large area of abnormal atrial wall which allows the development of electrical circus pathways. It is interesting that in other patients the arrhythmia was abolished by aneurysmectomy⁵ as in this patient.

Furthermore, aneurysmectomy has, in all the cases reported, resulted in the abolition of systemic emboli.⁶

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