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Brief Report

Intrapericardial giant left atrial appendage

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Abstract Contrary to what the name implies, the left atrial appendage is increasingly becoming an organ of clinical importance. Its role in the pathogenesis of thromboembolism is now well established. Enlargement of the appendage due to haemodynamic disturbances or anatomic weakness increases this risk, along with the potential for arrhythmia. We report a rare case of gross enlargement of the left atrial appendage within the pericardial cavity.

Keywords: Aneurysmal; dilatation

CONGENITAL INTRAPERICARDIAL ANEURYSMAL dilatation of the left atrial appendage is extremely rare, with hardly any reports in patients under 5 years of age. Differentiation from aneurysmal dilatation of the appendage secondary to herniation through a defect in the pericardial sac is often difficult. Even in asymptomatic patients, there is a potential for systemic embolisation and supraventricular dysrhythmia. We report a child aged less than 5 years in whom we found a giant left atrial appendage in the absence of any pericardial defect.

Case report

A boy, aged 4.5 years, was referred to our unit for further assessment subsequent to the clinical finding of a systolic murmur. His mother reported that he was more frequently "chesty", and tired on exertion, compared to his siblings. There was no history of asthma, palpitations, chest pains or syncope.

Clinical examination revealed a soft ejection systolic murmur in the pulmonary area. The electrocardiogram showed sinus rhythm with right bundle branch block. The chest X-ray was striking, with borderline cardiomegaly, and an abnormal left border to the cardiac shadow that was "shouldered" in the region of the left atrial appendage (Fig. 1a). The transthoracic echocardiographic findings were more revealing (Fig. 2). This showed a normally positioned left atrium behind the aorta, but with gross dilatation of the appendage. The mitral valve was normal, with no regurgitation. Subsequent magnetic resonance imaging (Fig. 1b) and cardiac catheterisation, confirmed that the left appendage was dilated, and projected parallel to the anterolateral surface of the left ventricle. There was suspicion of herniation through a defect in the pericardium, and surgery was undertaken with a view to repair the pericardial defect. The surgical approach was through a median sternotomy, but no pericardial defect was found. The left atrial appendage was broad-based, and continuous with the atrioventricular groove and the roof of the left atrium. The pectinate muscles seemed to continue down to the atrioventricular groove, contrary to the normal morphology, albeit that the tip of the appendage did not resemble the usual arrangement of a morphologically right appendage. There was no tendency to arrhythmia, and the appendage was normally contractile. Pulmonary veins were seen to discharge into the base of the left atrium. As the patient was asymptomatic, with no evidence of complications, we decided not to amputate or reduce the size of the appendage. The patient made an uneventful recovery, and continues to remain well.

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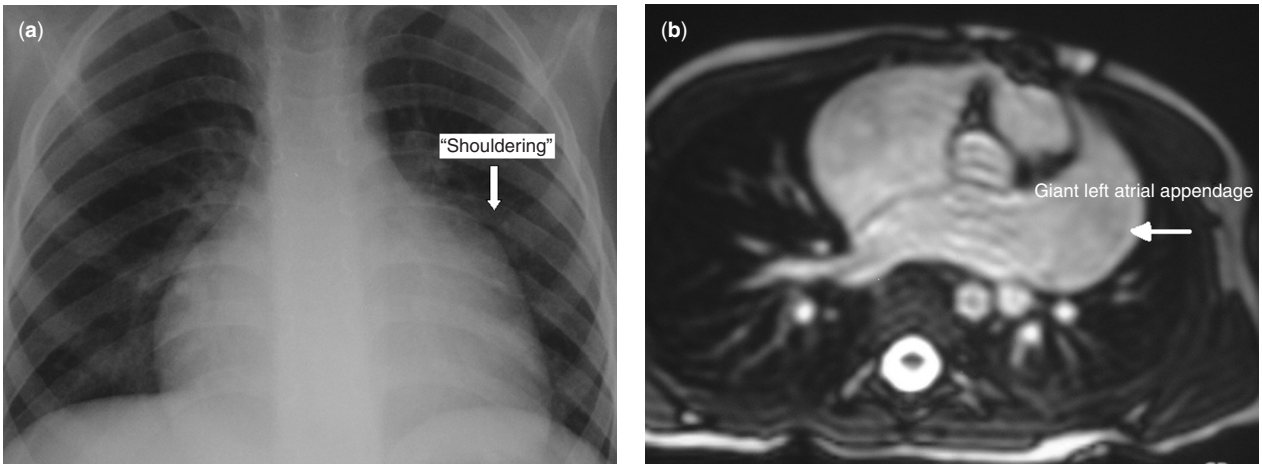


Figure 1.
*The chest X-ray (**a**) shows the "shouldering" of the left heart boarder, which is revealed by resonance imaging (**b**) to be produced by the aneurysmal left atrial appendage.*

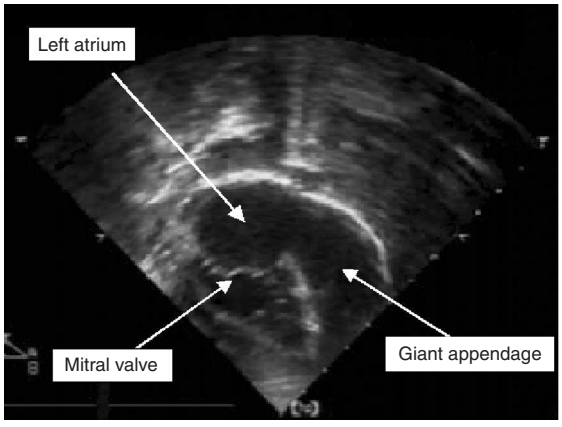


Figure 2.
Transthoracic echocardiogram confirms the gross enlargement of the left atrial appendage.

Discussion

Much literature has accumulated over the past few decades describing the clinical, radiological, echocardiographic and morphologic features of aneurysms of the atrial chambers. Left atrial enlargement reaching gigantic proportions can be either acquired or congenital. Although both are rare, acquired aneurysms are relatively more common, and have been associated with conditions with chronically increased atrial pressure, or those which weaken the atrial wall, such as rheumatic mitral valvar disease, tuberculosis and syphilitic myocarditis. Marfan's syndrome can also be a substrate for gross enlargement of the left atrium, even in infancy.[1]

Dilatation confined to the left atrial appendage, however, is unusual. To our knowledge, there are about 50 reported cases. Williams[2] described two varieties of congenital origin, one group occurring with intact pericardium, and the other in association with a pericardial defect. In the extra-pericardial variety, the appendage herniates through a defect in the pericardial sac, with the potential for incomplete emptying of the herniated portion in systole through the narrow "neck". This entity is relatively more frequent. Congenital intrapericardial aneurysmal dilatation of the appendage suggests a potential developmental weakness in the atrial wall.

Only one-third of the patients with congenital aneurysmal dilatation of the left atrium presents symptomatically before the age of 10 years. The average age at presentation is 26 years, with a marginal male preponderance.[3] Very few cases of an isolated giant left appendage with an intact pericardium have been described in patients as young as ours. Unsurprisingly, these patients remain asymptomatic as children, and clinical suspicion is usually aroused from an abnormal cardiac silhouette observed incidentally from a chest X-ray.

Dilatation of the left atrial appendage produces a bulge along the left border of the heart as viewed in the frontal projection, the bulge seen between the curves produced by the pulmonary trunk and the left ventricle. In patients with a narrow anteroposterior thoracic diameter, the heart is flattened between the sternum and the spine, causing the appendage to protrude from the left cardiac border. The differential diagnosis in such a setting would include partial absence of the pericardium, with herniation of the appendage through the defect, paracardiac tumours or cysts, enlargement of the coronary sinus, and aneurysm of the left ventricle. It was the abnormal left cardiac border in the chest X-ray that aroused our suspicion.

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Various invasive and non-invasive modalities have been used over the years for accurate preoperative diagnosis, including M-mode, cross-sectional and transoesophageal echocardiography, computerised tomography, magnetic resonance imaging and nuclear imaging.

Enlargement of the appendage can usually be detected accurately using M-mode echocardiography, but can be mistaken for significant pericardial effusion if not combined with cross-sectional imaging to provide accurate spatial orientation. The enlarged appendage is seen as a sonolucent space adjacent to the atrium in an off-axis apical four-chamber view. A transoesophageal scan may sometimes be needed to demonstrate the definite point of attachment to the heart. Theoretically, angiography could produce false negative images if the aneurysm is filled with thrombus.[4,5]

The electrocardiogram usually shows left atrial abnormalities, with broad and notched P waves. Supraventricular dysrhythmias are common, typically in the form of paroxysmal atrial tachycardia, atrial flutter, and atrial fibrillation. There is a strikingly high incidence of abnormalities of rhythm and systemic embolisation. About two-fifths of the patients experience atrial fibrillation, and one-fifth suffer strokes.[3] Chest pain can also occur, with or without bouts of tachycardia.

Excision of the aneurysmal appendage has been the therapeutic option in the majority of the reported cases. In our case, the absence of symptoms prompted us to avoid excision. There remains an argument, nonetheless, for surgical intervention to eliminate a potential source of systemic embolisation and supraventricular dysrhythmias. Time will tell whether these develop in our patient, but we hope not.

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