Congenital Giant Intrapericardial Aneurysm of Left Atrial Appendage: A Very Rare but Potentially Dangerous Entity

A. A. Rafighdoust, MD¹, A. Hamedanchi, MD¹ and A. Mirzaei, MD²

Abstract

A case of left atrial appendage (LAA) aneurysm misdiagnosed previously as partial pericardial defect is presented. This previously totally asymptomatic young multiparous woman was referred to us because of intermittent sharp chest pain and an abnormal CXR. Complementary imaging modalities revealed a large mass behind the left ventricle causing anterolateral hypokinesis. Although relatively symptomless, because of potential hazardous complications such as paroxysmal supraventricular tachycardia, syncope, sudden cardiac death, embolic phenomena and strangulation, the patient's LAA aneurysm was resected shortly after the confirmation of diagnosis. The postoperative course has been uneventful, and she is currently asymptomatic (*Iranian Heart Journal 2003; 4 (4):75-78).*

Congenital aneurysm of the left atrial appendage (LAA) is a very rare cardiac anomaly. Only 48 cases had been reported in the literature up to 1996,² with the vast majority presenting in otherwise healthy young patients. Informative data about these anomalies are lacking due to scarcity and rarity of the entity. These anomalies can present clinically with persistent arrhythmias and progress to stroke if not diagnosed and treated.

Our case is a 34-year-old woman diagnosed with a giant left atrial aneurysm presented with intermittent who palpitations and atypical angina. The only diagnostic clue was an abnormal chest Despite radiograph. the relatively symptomless condition, the patient underwent a successful surgical resection with cardiopulmonary bypass (CPB). The patient has been asymptomatic over the 13 months of follow-up ever since.

Case report

A 32-year-old woman was referred to us for the evaluation of atypical angina and an

abnormal chest radiograph. She had given birth to five normal children over the previous ten years and had been entirely symptom- free until several weeks before examination when she experienced sharp short-lasting chest pain together with palpitation. The history was otherwise for other unremarkable pertaining symptoms. Physical examination was normal except for a mid-systolic click. Electrocardiography was within normal limits. Chest radiography (Fig. 1) showed



Fig. 1. Chest radiograph depicting enlarged left cardiac border.

Correspondence to: A. A. Rafighdoust, MD, Department of Cardiology, Imam Reza Hospital, Mashhad University of Medical Sciences, WW, SID. ir Mashhad, Iran Tel: 0511-8593039

an enlarged left cardiac silhouette with a marked prominence of the upper and middle left heart border.

Partial pericardial defect with herniation of the LAA was diagnosed following transthoracic and transesophageal echocardiography (Figs. 2-4).



Figs. 2-4. TTE views suggesting pericardial defect and large herniated left atrial appendage.

Cardiac dysfunction caused by compression from the enlarged LAA was also noted. CT scan of the thorax added nothing to the entire clinical picture. Diagnostic right and left-sided catheterization was performed, and the levophase of main pulmonary artery (Fig. confirmed our erroneous nearly 5) echocardiographic diagnosis of partial pericardial defect.



Fig. 5. Levophase angiography view of pulmonary artery demonstrating large left atrium.

Chest pain, risk of future thromboembolic events, tachyarrhythmias, cardiac dysfunction, sudden cardiac death as well as potential hazard of strangulation supported rapid surgical strongly intervention for the patient. The intraoperative diagnosis (Figs. 6-8) was congenital aneurysm of LAA.



Fig. 6.



Figs. 6-8. Intraoperative views confirming large congenital left atrial appendage aneurysm.

Although differentiation between these two rare anomalies is reported preoperatively, we were unable to do so and the patient made an uneventful recovery and has remained asymptomatic for 13 months ever since.

Discussion

Giant intrapericardial aneurysms of the left atrial appendage and body are unusual cardiac anomalies.¹⁻³ They usually develop congenitally and have a risk of lifethreatening complications. Here, we report a case of an aneurysm of the left atrium that was incidentally found in a 32-yearold woman. The only important finding before echocardiography was an abnormal chest radiograph, stressing the utmost role of imaging in the clinical evaluation of

some complex forms of intermittent chest pain. The chest radiography at this time demonstrated a prominent left cardiac shadow suggesting LA enlargement, localized pericardial defect or some rare case of pericardial effusion. In addition to compression and indentation of the anterolateral wall of the left ventricle, hypokinesis was also noted in our patient, which has been reported before.⁴ This abnormality improved after surgical resection.

Victor attributed the aneurysm to congenital dysplasia of the "musculi pectinati" in the left atrial appendage and of the bands of atrial muscle from which they arise.⁵ In addition, they observed that the strap-like arrangements of musculi in both atria connected the outer band with the para-annular segment of the inner band. Utilizing intraoperative transesophageal echocardiography and surgical stapling devices, Burke et al. have streamlined their operative technique, allowing resection of a left atrial appendage aneurysm without cardiopulmonary bypass.⁶

Our patient had no important symptom or sign, but undoubtedly benefited from resection. Furthermore. surgical the thoracic pain mimicking pericarditis or angina associated with an abnormal chest X-ray must lead to cardiac sonographic study. In our patient, partial pericardial defect, a closely-related clinical and pathological entity, was firstly deduced, but did not interfere with our management strategy. This case. although rare. demonstrated the role of complementary imaging techniques in the recognition of rare but potentially hazardous diseases.

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