N Awasthy, M Tomar, S Radhakrishnan, S Shrivastava, and KS Iyer. Symptomatic giant left atrial aneurysm in a child: a rare entity. Images Paediatr Cardiol. 2010 Jan-Mar; 12(1): 9–13.

IMAGES in PAEDIATRIC CARDIOLOGY

Images Paediatr Cardiol. 2010 Jan-Mar; 12(1): 9–13.

PMCID: PMC3228329

Symptomatic giant left atrial aneurysm in a child: a rare entity

N Awasthy, M Tomar, S Radhakrishnan, S Shrivastava, and KS Iyer
Department of Pediatric Cardiology, Escorts Heart Institute and Reseach Center (EHIRC), Okhla road, New Delhi, 110025, India.
Corresponding author: Neeraj Awasthy, 123, Anand Kunj, Vikaspuri, New Delhi, India, 110018 ; Email: n_awasthy@yahoo.com

Abstract

Isolated left atrial aneurysms are rare entities in clinical practice. Usually the condition is diagnosed in the second to fourth decades of life. The presence of such lesions in the pediatric age group is scantily described. We present a 2 year boy who presented with complaints of excessive irritability, respiratory distress and swelling of the feet. On examination, child was tachypnoeic with irregularly irregular rhythm. Echo showed a huge aneurysmal LA appendage with severe left ventricle dysfunction. The child underwent surgical resection for same. Findings were confirmed intraoperatively but he continued to have low cardiac output state after the surgery, with frequent arrhythmias and expired on day 7 of surgery. The case is reviewed and compared with the available English literature.
MeSH: Heart defects, congenital, Left atrium, Aneurysm

Introduction

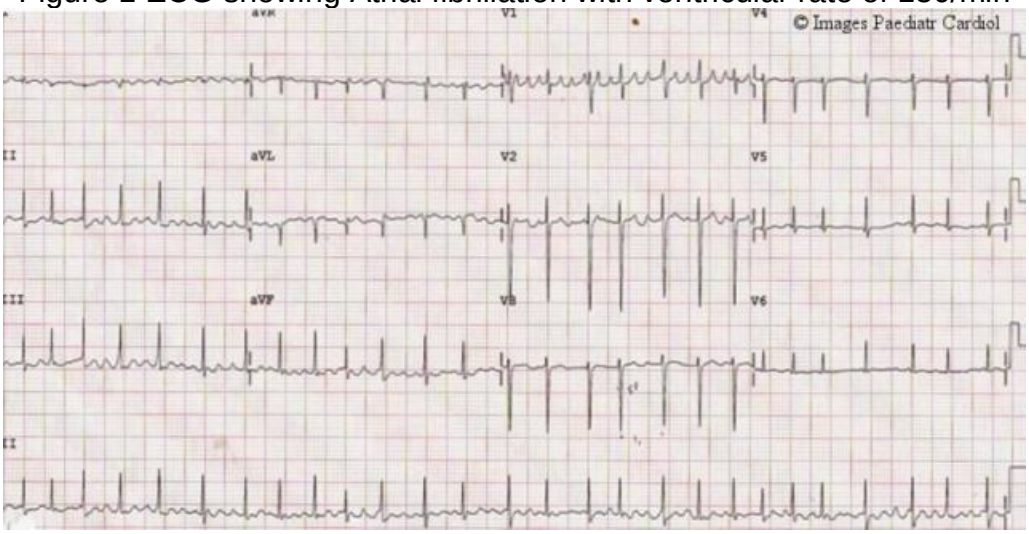
Aneurysm arising from the left atrial appendage is a rare entity in the pediatric population especially in the absence of associated anomalies. We present such a patient who presented in gross congestive heart failure with atrial fibrillation. Excision of the diverticulum was done under cardiopulmonary bypass. Our experience on this rare congenital disease is presented along with a review of the literature.

N Awasthy, M Tomar, S Radhakrishnan, S Shrivastava, and KS Iyer. Symptomatic giant left atrial aneurysm in a child: a rare entity. Images Paediatr Cardiol. 2010 Jan-Mar; 12(1): 9–13.

Patient

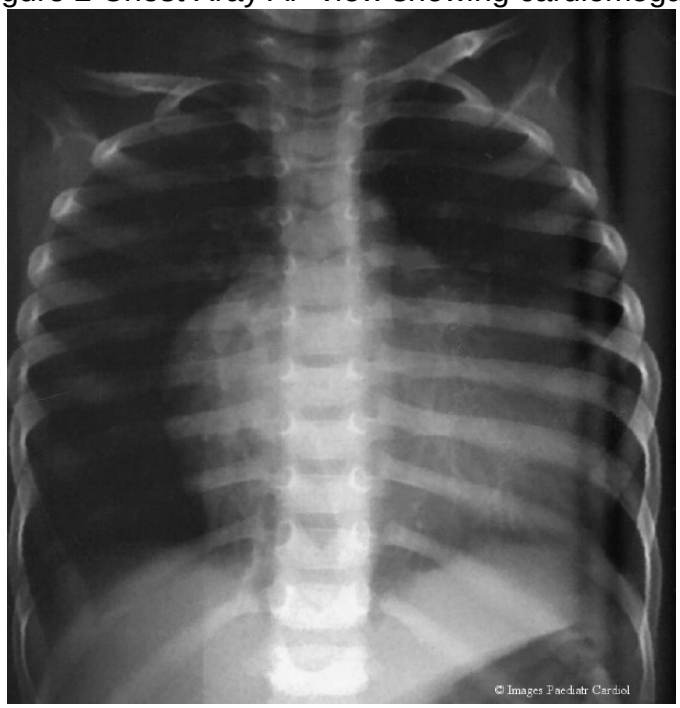
PA, a 2 year old boy presented with complaints of excessive irritability, respiratory distress for 15 days and swelling of the feet for 5 days. At the time of evaluation the child was in a stage of poor peripheral perfusion with cold clammy extremities and feeble peripheral pulses. There was tachypnea, tachycardia with irregularly irregular rhythm and pedal edema. ECG showed atrial fibrillation, with a ventricular rate of 150/ min (figure 1).

Figure 1 ECG showing Atrial fibrillation with ventricular rate of 150/min



Chest x-ray (posteroantrior view) showed cardiomegaly with a prominent left cardiac border (cardiothoracic ratio of 71% - figure 2).

Figure 2 Chest Xray AP view showing cardiomegaly



Echocardiography was done with Hewlett Packard 7500 with broad band transducers (S12,S8). Imaging from subcostal, parasternal long axis and short axis, and apical four chamber views showed concordant atrioventricular and ventriculoarterial connections, no septal defect, normal AV valves and semilunar valves with normal inflow velocities. There was severe LV dysfunction (LVEF 25%). In addition, there was an echo free space communicating with the left atrial appendage with evidence of spontaneous echo contrast. Color flow mapping confirmed its continuity with the left atrial appendage. Origin and flow in both left and right coronaries was normal (figures 3,4).

Figure 3 Modified Apical view showing large LA aneurysm

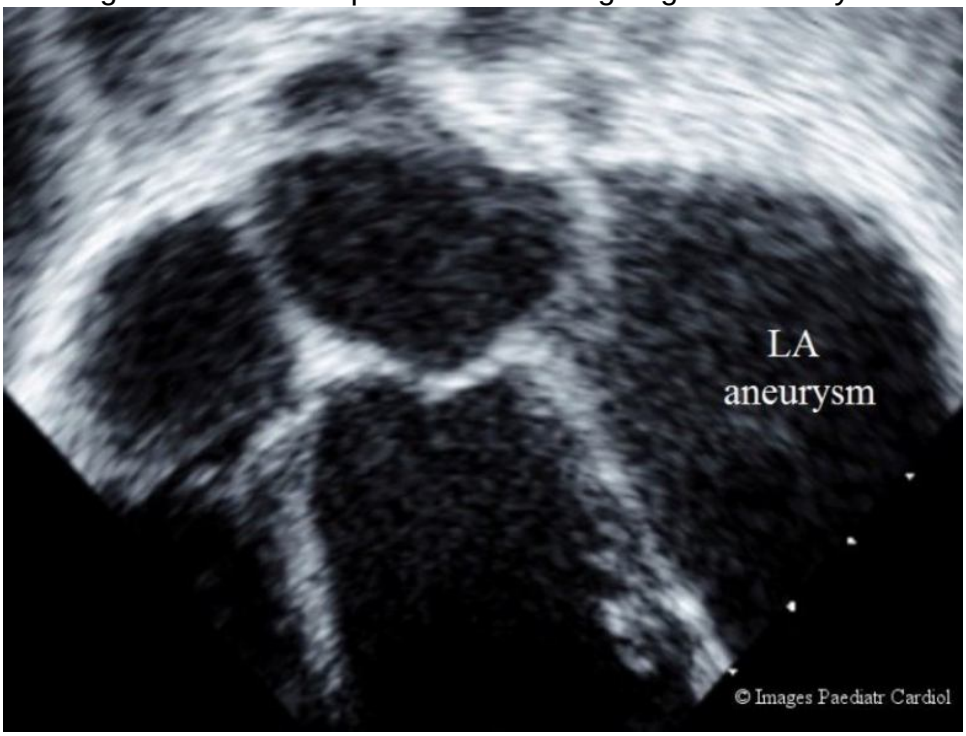
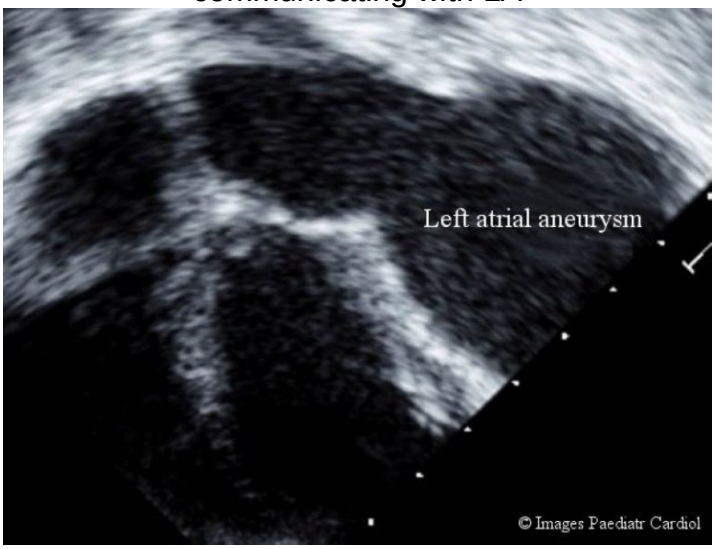
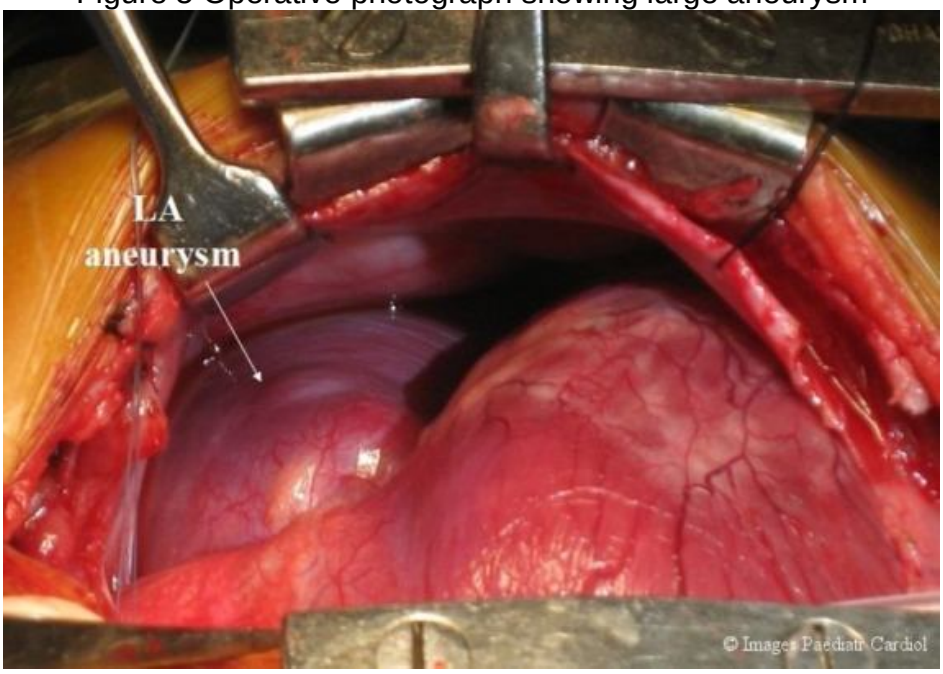


Figure 4 Modified Parasternal long axis view showing large LA aneurysm communicating with LA



The child was admitted in intensive care unit. He was started on intravenous diuretics (frusemide), inotropes (dobutamine and milrinone), and antiarrhythmic drugs (IV amiodarone). After two days of amiodarone, the rhythm reverted to sinus. With the probability of tachycardiomyopathy as a cause of ventricular dysfunction, he was taken up for surgical resection of the aneurysm.
Operative findings were consistent with the echocardiography findings (figure 5).

Figure 5 Operative photograph showing large aneurysm



Resection of the aneurysm was done under cardioplegia. The pathology report showed flattened tissue pieces with focally attenuated endocardium, and dilated intramural blood vessels, with mononuclear cell aggregates. There were no Ascoff nodules were seen. He had a difficult postoperative course with low output cardiac state, ejection fraction of 15%. Although the baseline rhythm reverted to sinus, there were frequent episodes of ventricular tachycardia requiring DC shock. Repeat echocardiography showed no improvement in LV EF, spontaneous echo contrast in LV and evidence of thrombus in RA. Child was continued on IV Amiodarone and heparin. The child continued to be in low cardiac output stage despite full ionotropic support and finally succumbed on the 9th postoperative day.

Discussion

Left atrial aneurysm is rare entity particularly in the pediatric population. The condition generally manifests itself in the second to fourth decades of life; however it can be diagnosed in newborns, infants or by fetal echocardiography.[1] The majority of patients are asymptomatic, but symptomatic adult patients usually present with palpitation, dyspnea on exertion and stroke, while in infants or children the aneurysm can be the

cause of cardiac arrest, respiratory distress, or cardiac tamponade.[2,3] Dilatation of the left atrial appendage is usually associated with mitral valve disease or herniation through a pericardial defect, but in the absence of a known predisposing factor the condition has been assumed to be of congenital origin.[4] Victor and Nayak postulated that the cause of the aneurysm may be due to congenital dysplasia of the pectinate muscles and of the left atrial muscle bundles related to them. Such dysplasia would impair contraction of the appendage during atrial systole.[4]

A chest x-ray showing a prominent left cardiac border should raise the suspension of a left atrial aneurysm. Echocardiography including transesophageal echocardiography is the most useful noninvasive means of study. Magnetic resonance imaging is also helpful for establishing the diagnosis and delineating the relation to the surrounding structures with the use of coronal imaging plane. In the past cardiac angiography was essential for confirming the diagnosis.[5]

The complications associated with this abnormality can be devastating, especially thromboembolism and cardiac arrhythmia; for these reasons, surgical resection is recommended even in asymptomatic cases. Though the left atrial appendage aneurysm is a rare entity presenting in older age group, it can have presentation as refractory arrhythmia leading to severe LV dysfunction. It is important thus to make a timely diagnosis to have good outcome.[6]

References

1. Fountain-Dummer RR, Wiles HB, Shuler CO, Bralley SM, Shirali GS. Recognition of left atrial aneurysm by fetal echocardiography. Circulation. 2000;102:2282–83.[PubMed: 11056106]

2. Park JS, Lee DH, Han SS, Kim MJ, Shin DG, Kim YJ, Shim BS. Incidentally found, growing congenital aneurysm of left atrium. J Korean Med Sci. 2003;18:262–66. [PMCID: PMC3055018][PubMed: 12692426]

3. Morales JM, Patel SG, Jackson JH, Duff JA, Simpson JW. Left atrial aneurysm. Ann Thorac Surg. 2001;71:719–22.[PubMed: 11235743]

4. Victor S, Nayak V. Aneurysm of the left atrial appendage. Tex Heart Inst J. 2001;28:111–18. [PMCID: PMC101149][PubMed: 11453121]

5. LaBarre T, Stamato N, Hwang M, Jacobs W, Stephanides L, Scanlon P. Left atrial appendage aneurysm with associated anomalous pulmonary venous drainage. Am Heart J. 1987;114:1243–45.[PubMed: 3673894]

6. Burke RP, Mark JB, Collins JJ, Jr, Cohen LH. Improved surgical approach to left atrial appendage aneurysm. J Cardiac Surg. 1992;7:104–7.