Congenital aneurysm of both left ventricle and left atrium

Ryan F Halas, Christopher J Schmehil[1], Gary R Ten Eyck[2], James L Loker[3]

Department of Internal Medicine-Pediatrics, [1]Departments of Pediatrics and [2]Biomedical Sciences, Western Michigan University Homer Stryker M.D. School of Medicine, [3]Department of Pediatric Cardiology, Bronson Children's Hospital, Kalamazoo, MI, USA

ABSTRACT

This is a case of both congenital left ventricular (LV) free wall submitral aneurysm and left atrial appendage aneurysm with 6 years of clinical follow-up. Each lesion is a rare entity, and to the best of our knowledge, this is the first case in medical literature of both lesions occurring in the same patient, raising the likelihood of a common etiology. The workup was initiated in the third trimester of fetal life with irregular heart rate and abnormal fetal ultrasound and echocardiogram at that time. The patient required emergent atrial appendage plication due to blood clot formation and suffered from multiple other complications including ventricular ectopy and surgically induced pseudoaneurysm. Follow-up interval echocardiograms have revealed continued good LV function with persistent LV aneurysm. In review of the case, there were several potential *in utero* causes including maternal viral upper respiratory infection and bacteriuria with exposure to amoxicillin. These as well as other considerations are discussed along with a brief review of these rare lesions, usual presentation, and known associations.

Keywords: Congenital left atrial appendage aneurysm, congenital left ventricular wall aneurysm, left atrial appendage aneurysm, left ventricle aneurysm, left ventricle free wall aneurysm

INTRODUCTION

The heart is one of the earliest organs to develop in embryologic life and is susceptible to insult leading to a myriad of different congenital heart defects including congenital aneurysms. Aneurysm is defined as an outpouching which contains all histologic components of the tissue wall, whereas a pseudoaneurysm is a contained rupture of some components of a histologic wall. Congenital left ventricular (LV) aneurysm and diverticulum are rarely encountered congenital defects thought to originate around the fourth embryonic week.[1] Congenital atrial aneurysm is a rarer defect with <80 cases described before 2007.[2] We present the first reported co-occurrence of both congenital LV and left atrial (LA) appendage aneurysm with 6 years of clinical follow-up.

CASE REPORT

A 24-year-old G4P2 female presented for fetal arrhythmia at 33 weeks' gestation. Fetal ultrasound revealed enlarged heart with small periventricular effusion [Figure 1]. Detailed history revealed Group B Streptococcus (GBS) bacteriuria treated with a standard course of ampicillin between weeks 2 and 6 of gestation and upper respiratory infection at approximately 17–19 weeks gestation. The only other medication used was prenatal vitamins. Family history was negative for congenital heart disease or connective tissue disease. Pregnancy history was positive for a first trimester spontaneous abortion and two full-term vaginal deliveries, one of which was complicated by pregnancy-induced hypertension. A fetal echocardiogram revealed no arrhythmia with preserved ventricular function, and no LV aneurysm was appreciated at that time. She was followed with weekly nonstress tests, which remained normal. She did not have any further complications or exposures, and at 39 weeks' gestational age, she had an uncomplicated vaginal delivery of a 3.245 kg boy.

The infant continued to have irregular heart rate, and an electrocardiogram revealed frequent premature

Access this article online

Quick Response Code:

Website: www.annalspc.com

DOI: 10.4103/apc.APC_55_17

How to cite this article: Halas RF, Schmehil CJ, Ten Eyck GR, Loker JL. Congenital aneurysm of both left ventricle and left atrium. Ann Pediatr Card 2018;11:97-9.

Address for correspondence: Dr. Ryan F Halas, DO, 1000 Oakland Dr, Kalamazoo, MI 49008, USA. E-mail: ryan.halas@med.wmich.edu

ventricular contractions but no runs of ventricular tachycardia. Vital signs were normal and physical examination noted a 2/6 systolic ejection murmur loudest at the left midsternal border with some radiation to the back. An echocardiogram revealed the presence of large LA appendage aneurysm and large LV free wall aneurysm with good biventricular function. He was started on Inderal and aspirin and was observed in the neonatal Intensive Care Unit with stable clinical course before repeat echocardiogram at 1 week of age showed the development of a large blood clot in the LA appendage. He was placed on enoxaparin sodium (Brand name - Lovenox, Manufacturer - Sanofi-aventis) and was transferred to a quaternary facility [Figure 2].

Cardiac magnetic resonance imaging confirmed LA appendage aneurysm with blood clot and large left free wall LV aneurysm. Surgical LA aneurysm resection was performed, but the clot had already embolized to the left iliac bifurcation without evidence of limb ischemia [Figure 3].

His initial postoperative course was uncomplicated, and he was discharged on Coumadin therapy. Postoperative echocardiogram revealed the development of a left ventricle apex pseudoaneurysm which required surgical plication [Figure 4].

Coagulation workup was negative, and Coumadin was discontinued with continuation of aspirin therapy. Serial echocardiograms continue to show large aneurysm of the left ventricle free wall with good LV function [Figure 5]. The ventricular ectopy decreased, and beta-blockers were discontinued until the age of six when he developed dizziness and ventricular ectopy.

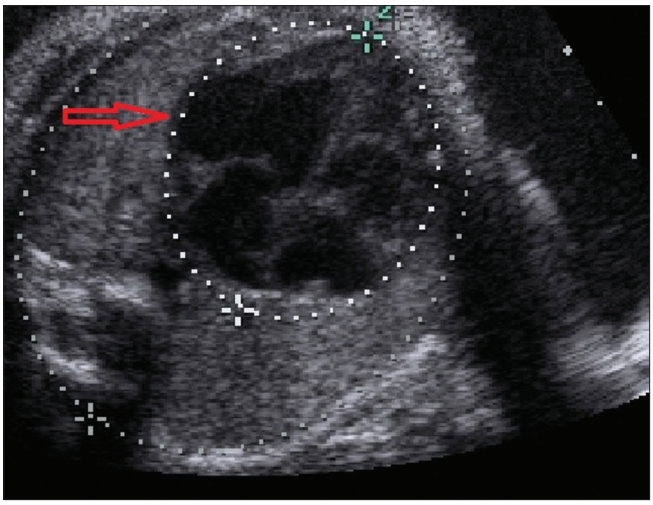

Figure 1: Prenatal US at 33 weeks' gestational age with red arrow showing dilation of the left ventricle directly under the mitral valve. Given echocardiogram and magnetic resonance imaging in the 1st week of life, this was likely aneurysmal dilation. The left atrial aneurysm is not visualized here or during prenatal US

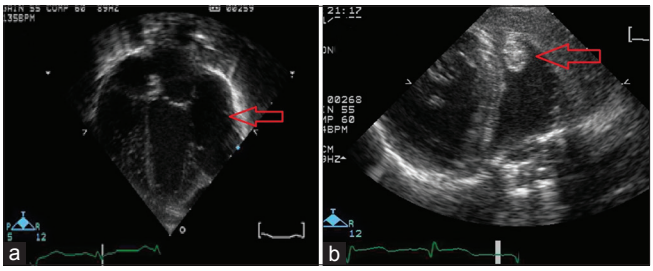

Figure 2: Echocardiogram imaging: (a) Four chamber view, day 1 of life, showing large left ventricle submitral free wall aneurysm. (b) Parasternal short axis view, day 7 of life, showing left atrial aneurysm with hyperechoic mass at apex (arrow), presumed to be a blood clot

DISCUSSION

This is the first reported co-occurrence of congenital LV free wall aneurysm and congenital LA appendage aneurysm, to the best of our knowledge. Each lesion is associated with multiple other cardiac defects, but this combination has never been reported.[1] It is commonly thought that LV aneurysms occur due to developmental defects as early as the 4th week of gestational age, and there are multiple variations of proposed insults from defect of the cardiac jelly due to inadequate blood supply to defects in the muscular fibrous junction below the intermediate portion of the posterior mitral leaflet.[3-5] None of these proposed mechanisms would fully account for an aneurysm in the ventricular and atrial wall. Even less is known about etiologies of congenital LA appendage aneurysm. It is suggested that these are the result of dysplasia of the LA muscle.[6,7]

It is difficult to implicate a single embryological etiology for these two aneurysms. This is in part because extrinsic damage to the left heart before atrioventricular septation

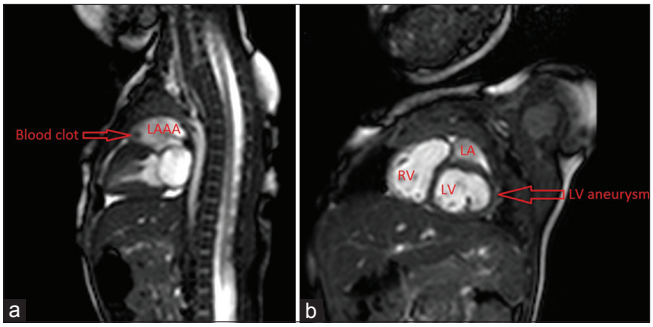

Figure 3: (a) Sagittal plane T2 cardiac magnetic resonance imaging, day 7 of life, showing left atrial appendage aneurysm and hypoechoic blood clot at the ventral aspect. The left ventricular free wall aneurysm can be seen directly below left atrial. (b) Sagittal oblique cardiac magnetic resonance imaging with free wall left ventricular aneurysm with free communication to the left ventricle

would likely be catastrophic. It is well known that there is a signal cascade of homeobox genes (PITX2) and transcriptional factors that are responsible for normal left side development although the atrioventricular

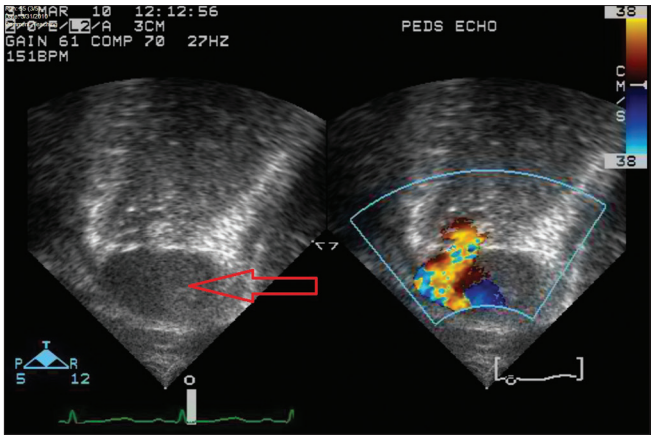

Figure 4: Postoperative apical view echocardiogram with and without Doppler flow showing the development of new left ventricular apical pseudoaneurysm. Arrow shows apical pseudoaneurysm and color flow study shows flow into the pseudoaneurysm during cardiac systole

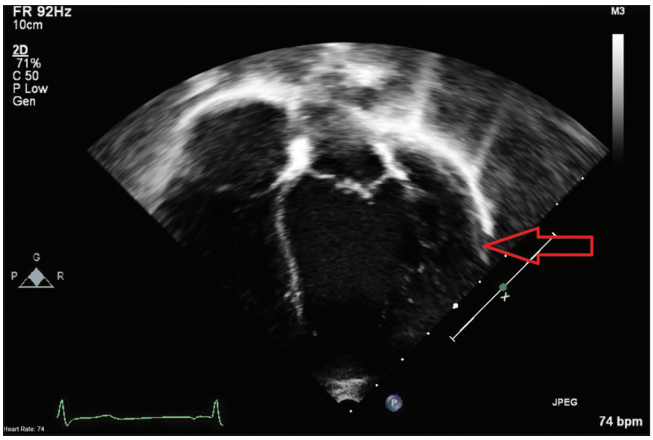

Figure 5: Apical 4 chamber view echocardiogram performed at 6 years of age showing persistent large submitral left ventricular aneurysm marked by red arrow

development in our patient appeared normal.[8] It seems that any significant defect of adhesion factors or connective tissue would result in more global effect on the heart although the later development of a pseudoaneurysm after surgery may lend more credence to this possible etiology. We hypothesize that the most likely unifying etiology remains acquired disease during fetal life.

The patient had two possible insults during fetal life – GBS bacteriuria between weeks 2 and 4 in gestation which was treated with ampicillin and a presumed viral upper respiratory infection at approximate gestational age between 17 and 19 weeks. Our patient's GBS bacteriuria and ampicillin therapy occurred at a time during fetal development in which LV aneurysm is thought to form, but neither GBS bacteriuria nor ampicillin have any known teratogenic or fetal cardiotoxic effects. Fetal cardiac insult from maternal viral infection seems to be a more likely culprit, but the timing of her symptoms was early in the second trimester, after most of cardiac development. It is possible that viral myocardial damage leads to both ventricular and atrial wall weakness and subsequent aneurysmal dilation along with ventricular ectopy.

CONCLUSION

This is possibly the first reported case of co-occurrence of both LA and LV true congenital aneurysms. Although there are multiple proposed mechanisms of both congenital ventricular aneurysm and congenital atrial appendage aneurysm, it appears that most of these models would not account for the formation of both lesions. In our patient, we feel that the most likely cause was an acquired fetal cardiac disease from GBS urinary tract infection with ampicillin exposure in the first trimester or viral cardiomyopathy during the second trimester. In the future, it will be important to investigate similar cases for commonality leading to a more accurate assessment of possible etiology. The patient is fortunate not to have had any severe complications related to these malformations but continues to have a large ventricular aneurysm and some ventricular ectopy which will require thorough follow-up.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Ohlow MA. Congenital left ventricular aneurysms and diverticula: Definition, pathophysiology, clinical relevance and treatment. Cardiology 2006;106:63-72.
2. Chowdhury UK, Seth S, Govindappa R, Jagia P, Malhotra P. Congenital left atrial appendage aneurysm: A case report and brief review of literature. Heart Lung Circ 2009;18:412-6.
3. Kontozis L, Skoularigis J, Skudicky D, Sareli P. Submitral aneurysm. Circulation 1998;98:1698.
4. Abrahams DG, Barton CJ, Cockshott WP, Edington GM, Weaver EJ. Annular subvalvular left ventricular aneurysms. Q J Med 1962;31:345-60.
5. Chesler E, Joffe N, Schamroth L, Meyers A. Annular subvalvular left ventricular aneurysms in the South African Bantu. Circulation 1965;32:43-51.
6. Victor S, Nayak VM. Aneurysm of the left atrial appendage. Tex Heart Inst J 2001;28:111-8.
7. Pomerantzeff PM, Freyre HM, de Almeida Brandão CM, Pereira Barreto AC, Almeida de Oliveira S. Aneurysm of the left atrial appendage. Ann Thorac Surg 2002;73:1981-3.
8. Sadler TW. Langman's Medical Embryology. 15th ed. Baltimore, MD: Lippincott Williams & Wilkins, A Wolters Kluwer Business; 2015.