

Congenital pseudoaneurysm of mitral-aortic intervalvular fibrosa masquerading as left atrial mass in fetal life

Shanthi Chidambarathanu, Vijayalakshmi Raja, Indrani Suresh

Fetal Cardiology Unit and Pediatric Cardiac Services, Mediscan Systems, Chennai, Tamil Nadu, India

ABSTRACT

A 28-week-old fetus was detected to have a single left atrial mass in prenatal ultrasound. Postnatal echocardiography showed an aneurysm between the anterior mitral leaflet and aortic valve, to the left of atrioventricular junction and communicating with the left ventricle through a narrow mouth. It probably originated from the mitral-aortic intervalvular fibrous tissue and an inherent weakness at this site might be the cause. Reported cases of pseudoaneurysm of mitral-aortic intervalvular fibrosa and subvalvular ventricular aneurysms seen following infective endocarditis, surgery, or trauma seem to have a similar anatomical background. This case explains the possibility of congenital aneurysm in this location which needs to be considered a differential diagnosis in similar cases.

Keywords: Aorto-mitral valvular curtain, fetal echocardiography, left atrial aneurysm, left atrial mass, mitral-aortic intervalvular fibrosa

INTRODUCTION

Prenatal detection of intracardiac masses has increased in recent years with advancement in technology and expertise. We describe in this report an unusual cardiac defect which presented as an intracardiac mass during third-trimester obstetric imaging.

CASE REPORT

A primigravida mother was referred at 28 weeks of gestation for fetal echocardiography following suspicion of a left atrial rhabdomyoma. Fetal echocardiography showed a single nonpedunculated mass located at the atrioventricular junction close to the primum atrial septum and the anterior leaflet of mitral valve [Figure 1a]. It measured 8 by 7 mm in size and had echogenic margins with central echolucency. Mitral valve looked normal and

flow from left atrium to left ventricle was unobstructed [Supplementary Video 1]. The mass could be visualized adjacent to the aortic valve also, with the anterior tilt of the probe [Figure 1b]. Cardiac rhythm was normal, and no other mass could be seen within the fetal heart. Fetal growth was within normal limits and rest of fetal parts looked normal. We considered it to be a left atrial mass of uncertain origin as the echogenicity and location were not typical of rhabdomyoma, myxoma, or fibroma, the tumors seen commonly in fetal heart and family was counseled accordingly. Review at 32 weeks did not show an increase in the size of the mass or any additional mass within the heart and cardiac flow pattern remained normal. Postnatal follow-up details were available only after 9 months of delivery. It was a girl baby, born by normal vaginal mode, weighed 3.2 kg and had an uneventful postnatal period. Postnatal echocardiogram had reported a left atrial mass and child had been on medical follow-up as she remained

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Address for correspondence: Dr. Shanthi Chidambarathanu, Mediscan Systems, No 197, Dr. Natesan Road, Mylapore, Chennai - 600 004, Tamil Nadu, India.
E-mail: shanthic2009@gmail.com

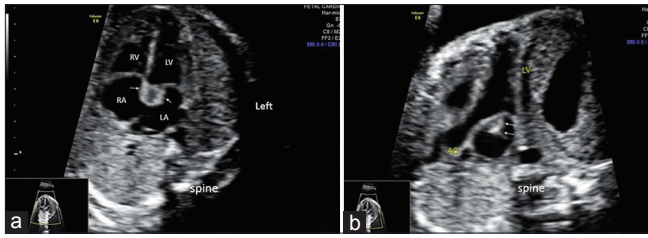


Figure 1: Fetal echocardiography images at 28 weeks of gestation: (a) Four-chamber view of fetal heart showing the mass with central echolucency seen at the atrioventricular junction above the anterior mitral leaflet (arrows), (b) same mass seen in relation to the aortic valve in the left ventricular outflow view in sagittal plane (arrows); RA: Right atrium, LA: Left atrium, RV: Right ventricle, LV: Left ventricle, AO: Aorta, spine-fetal spine, Left-left side of fetus

asymptomatic. During her visit to our department, she was a well-grown infant with normal external features and development. There was no cyanosis, pulses were normal, a nonejection click was heard over the left sternal border, and there was no murmur audible. Chest X-ray showed normal cardiothoracic ratio and lung fields. Transthoracic echocardiography revealed an aneurysmal pouch located anteromedial to the anterior mitral leaflet, posterior to the aortic valve, and close to the interatrial septum and seen projecting into the left atrial cavity [Figure 2a and b]. Wall of the aneurysm was redundant with flower-like projections. It was communicating with the left ventricular cavity through a narrow mouth of 3 mm in diameter, which could be demonstrated by color Doppler assessment [Figure 2c]. It was difficult to assess if the aneurysm was within the left atrial cavity or adjacent to it, but no evidence of communication to the left atrial cavity was noted [Supplementary Video 2]. Moreover, it was not obstructing the subaortic area. Both leaflets of the mitral valve and subvalvular apparatus looked normal. There was no other abnormality noted in the echocardiography and electrocardiogram showed normal rhythm. We revisited the fetal echocardiography video recordings, where blood flow from the left ventricle into the aneurysmal pouch could be appreciated in color Doppler in retrospect.

DISCUSSION

The aneurysm seen in this case seems similar to the reported cases of pseudoaneurysm of mitral-aortic intervalvular fibrosa (MAIVF), mostly seen following infective endocarditis.^[1-3] MAIVF or aorto-mitral valvular curtain is the tissue located in the fibrous connection between the aortic valve cusp and anterior leaflet of mitral valve.^[4,5] As perforation of this tissue during surgery or trauma is known to cause the aneurysm, an inherent deficiency at this site may explain the congenital aneurysm formation. Chesler *et al.* in 1982 and Barranhas *et al.* in 2012 have illustrated a type of subvalvular ventricular aneurysm occurring at the MAIVF site.^[6,7] Location of the defect seen in our case seems similar to

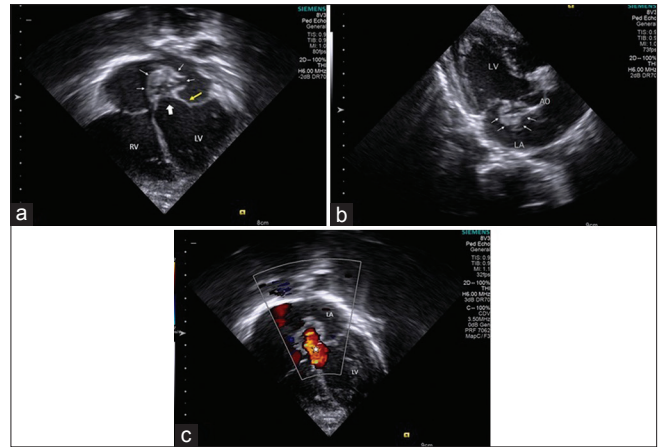


Figure 2: Postnatal transthoracic echocardiography images: (a) Apical four chamber view showing the aneurysm (small arrows) seen between the atrioventricular junction and the anterior mitral leaflet (yellow arrow); the mouth of the aneurysm connecting to left ventricle shown in broad arrow. (b) Parasternal long axis view showing the relationship of the aneurysm (small arrows) to the aortic valve and anterior mitral leaflet. (c) Apical four chamber view showing color flow inside the aneurysm (star) communicating with the left ventricle. LA: Left atrium, LV: Left ventricle, RV: Right ventricle, AO: Aorta

the above mentioned, but of congenital origin. So far, only two cases of congenital pseudoaneurysm at this site have been reported to the best of our knowledge. One in a 4-year-old child with Shone's anomaly and another in an adult.^[8,9] Our case seems to be the first isolated congenital aneurysm seen in infancy which mimicked a left atrial mass in prenatal imaging. Unusual location and color flow into the mass during prenatal echocardiography were the findings in favor of an aneurysm which were overlooked during fetal imaging, probably due to the rarity. Despite a preplanned delivery and postnatal care in a tertiary level unit, the actual diagnosis got delayed, which reflects on the need for awareness about this entity and improved coordination between fetal and postnatal units. The characteristic systolic expansion and diastolic collapse of the aneurysm seen in echocardiography might help in differentiating it from a mass. Indication for surgical repair is not clear in asymptomatic cases. Our patient is on medical follow-up with endocarditis prophylaxis as it is a small aneurysm without any hemodynamic compromise. Need for intervention and timing may be decided in future based on further evolution. Complications such as rupture, coronary artery compression, aortic or mitral valve disease, and thromboembolism noted rarely in the acquired aneurysms have to be kept in mind during follow-up.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Sudhakar S, Sewani A, Agrawal M, Uretsky BF. Pseudoaneurysm of the mitral-aortic intervalvular fibrosa (MAIVF): A comprehensive review. *J Am Soc Echocardiogr* 2010;23:1009-18.
2. Xie M, Li Y, Cheng TO, Wang X, Lu Q, He L, *et al.* Pseudoaneurysm of the mitral-aortic intervalvular fibrosa. *Int J Cardiol* 2013;166:2-7.
3. Sahan E, Gül M, Sahan S, Sokmen E, Guray YA, Tufekçioğlu O. Pseudoaneurysm of the mitral-aortic intervalvular fibrosa. A new comprehensive review. *Herz* 2015;40 Suppl 2:182-9.
4. Gross L, Kugel MA. Topographic anatomy and histology of the valves in the human heart. *Am J Pathol* 1931;7:445-74.
5. Anderson RH. Clinical anatomy of the aortic root. *Heart* 2000;84:670-3.
6. Chesler E, Mitha AS, Edwards JE. Congenital aneurysms adjacent to the annuli of the aortic and/or mitral valves. *Chest* 1982;82:334-7.
7. Barranhas AD, Dias MC, Dos Santos AA, Marchiori E, Nacif MS. Pseudoaneurysm of the mitral-aortic intervalvular fibrosa presenting after chest trauma and diagnosed by cardiac magnetic resonance: A case report. *J Med Case Rep* 2012;6:357.
8. He D, Sinha P, Olivieri L, Jonas RA. Congenital aneurysm of the aortomitral intervalvular fibrosa. *Ann Thorac Surg* 2015;99:314-6.
9. Garçon P, Lavie-Badie Y, Cador R, Bical O, Abbey-Toby A. An unreported congenital anomaly: Aneurysm of the lateral half of the mitral valve. *Echocardiography* 2015;32:1594-6.