Pseudoaneurysm of Mitral-Aortic Intervalvular Fibrosa Imitating a Rhabdomyoma in a Fetus

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INTRODUCTION

Pseudoaneurysm of mitral-aortic intervalvular fibrosa (P-MAIVF) is an uncommon finding arising from the fibrous layer of tissue between the mitral and aortic valves. It is typically associated with infections, such as endocarditis, and trauma from cardiac surgery.[1,2] Congenital presentations are extremely rare. Here we present a case of an infant with a prenatally diagnosed cardiac rhabdomyoma that was shown postnatally to be a P-MAIVF, underscoring the importance of keeping this diagnosis in consideration for atrial masses noted on fetal echocardiography.

CASE PRESENTATION

A 21-year-old woman was initially referred at 31-1/7 weeks' gestation for fetal echocardiography because of concern for a cardiac mass recognized during obstetric ultrasound. The first fetal echocardiogram demonstrated a large echogenic mass in the anterior left atrium measuring 8.1 × 4.3 mm (Figure 1A, Video 1). The mass was associated with the atrial septum near the crux of the heart. It was not near the inflow or outflow tract, and there was no evidence of valvar dysfunction or outflow tract obstruction. No flow by color Doppler was noted inside the mass at this time. There was normal biventricular systolic wall motion and normal cardiac rhythm, and no other masses were visualized. The mass was thought most consistent with an atrial rhabdomyoma because of its increased echogenicity. The mother was counseled on rhabdomyoma and its strong association with tuberous sclerosis. The mother was seen prenatally by neurology because of concern for tuberous sclerosis. Of note, the patient also reported a history of supraventricular tachycardia managed on metoprolol. The patient underwent follow-up fetal echocardiography at 37-6/7 weeks' gestation, which continued to demonstrate a large echogenic mass, measuring 8 × 10 mm at that time, in the flap of the septum primum, which was still thought to be most consistent with a rhabdomyoma. The fetal rhythm continued to be normal, and there was no obstruction of the inflow or outflow tract.

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The mother gave birth to a male infant at 39 weeks' gestation via vaginal delivery without complications. The infant weighed 3,295 g, Apgar scores were 7 and 9, and the infant received routine resuscitation. The infant was admitted to the neonatal intensive care unit after birth in the setting of the prenatal diagnosis of rhabdomyoma for monitoring for possible development of arrythmias and workup for tuberous sclerosis. The postnatal echocardiogram demonstrated an echogenic mass at the atrioventricular junction along the atrial septum measuring 7.4 × 9.3 mm (Figure 1B, Video 2). There was blood flow into the mass during systole and flow into the left ventricular outflow tract during diastole, consistent with a P-MAIVF (Figure 2, Video 3). No other intracardiac masses were seen. Follow-up echocardiography on the third day of life again demonstrated a P-MAIVF, with no change in size. The echocardiographic findings were otherwise normal for age, with no other congenital heart defects. Clinically, the infant did well and remained hemodynamically stable, with stable size of the pseudoaneurysm and no arrhythmias. The mother and infant were discharged home on the third day of life with outpatient cardiology follow-up. The infant continued to do well, without growth of the pseudoaneurysm, rupture, or obstruction. Daily aspirin for the infant was started because of concern for thrombosis, and the infant continues to be monitored by echocardiography.

DISCUSSION

The mitral-aortic intervalvular fibrosa is a delicate layer of avascular tissue between the aortic valve annulus and the anterior mitral valve leaflet that is more susceptible to infection and damage during cardiac surgery.[1] Pseudoaneurysm of the mitral-aortic intervalvular fibrosa remains clinically rare and is typically found in adults after infectious or traumatic insult.[2] In the pediatric population, congenital cases have been noted, along with its association with congenital heart disease, including septal defects, coarctation of the aorta, and bicuspid aortic valve.[2,3] A literature review of pediatric cases of P-MAIVF by Ahmed *et al.*[1] demonstrated 46 pediatric cases in total, with ages ranging from 0 to 21 years, with three of these being diagnosed as isolated findings on fetal echocardiography. Hogan *et al.*[3] also reported an additional five prenatally diagnosed cases of congenital P-MAIVF, with two cases being isolated findings without other cardiac anomalies. All fetal cases were noted to be atrial masses at the time of fetal echocardiography and had color flow into the lesion, raising suspicion for P-MAIVF.[3-6]

In the diagnosis of P-MAIVF, both the location of the mass and its connection to the left ventricular outflow and to-fro flow can be used to distinguish this diagnosis from other potential etiologies of an intracardiac mass, such as rhabdomyoma, other intracardiac tumors, and thrombus. Furthermore, in this case the mass was initially thought to be a rhabdomyoma, which is one of the most common fetal cardiac tumors, though only 30% of rhabdomyomas present as single

VIDEO HIGHLIGHTS

Video 1: Fetal two-dimensional echocardiography in the four-chamber view at 31 weeks' gestation demonstrating the P-MAIVF in the anterior left atrium associated with the septum near the crux measuring 8.1 × 4.3 mm.

Video 2: Two-dimensional transthoracic echocardiography in the apical four-chamber view on the first day of life demonstrating the P-MAIVF at the atrioventricular junction along the atrial septum, measuring 7.4 × 9.3 mm.

Video 3: Two-dimensional transthoracic echocardiography in the apical four-chamber view on the first day of life demonstrating to-fro flow in the P-MAIVF by color Doppler with blood flow into the mass during systole and flow into the left ventricular outflow tract during diastole.

Video 4: Fetal two-dimensional echocardiography in the four-chamber view at 31 weeks' gestation on the initial study demonstrating flow into the P-MAIVF in systole by color Doppler. This was not noted on initial review of the study.

Video 5: Fetal two-dimensional echocardiography in the four-chamber view on follow-up study at 37 weeks' gestation demonstrating flow into the P-MAIVF in systole by color Doppler.

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tumors.[7] Other characteristic findings of P-MAIVF include arising in the region of mitral-aortic continuity, expansion of the mass in systole and collapse in diastole, and the to-fro flow, which can be demonstrated by color and pulsed-wave Doppler.[1,5,6] In our case, color flow within the atrial mass was not initially noted on fetal echocardiography, but upon review of images, there was to-fro flow on both initial fetal echocardiography at 31 weeks' gestation and follow-up echocardiography at 37 weeks' gestation, which is more consistent with the diagnosis of P-MAIVF (Figure 3; Videos 4 and 5). Color flow within the atrial mass was better demonstrated prenatally when using higher Nyquist limits (50-90 cm/sec). Color flow into the P-MAIVF was more readily apparent postnatally, when higher frequency transducers allowed better resolution for both two dimensional and color flow mapping modalities. In fetal echocardiography, as in postnatal scanning, optimization of color flow mapping includes the optimal choice of image depth and sector width choice of the smallest color box appropriate to the area of interest and optimizing the angle of interrogation, Nyquist limit, and wall filters. Gain and compression settings were manipulated in prenatal and postnatal studies on an individual basis to enhance optimal quality imaging.

The natural history of congenital P-MAIVF is still uncertain, and there appear to be varying clinical courses depending on the complications present. Complications can include thrombosis, infection, rupture, and compression of adjacent structures.[1-3] Other imaging modalities, such as transesophageal echocardiography, cardiovascular computed tomography, and magnetic resonance, may be used postnatally to further diagnose and characterize the mass, including its relationship to other cardiac structures.[8,9] This can aid in potential surgical planning. In general, uncomplicated, asymptomatic patients are managed conservatively, whereas patients with symptoms and/or complications undergo surgical repair.[4,10] Our patient has remained asymptomatic and has been followed conservatively with echocardiography and clinical assessment. We agree with conservative management postnatally in the absence of other findings or clinical symptoms. The initiation of aspirin can be considered as thrombus prophylaxis in these patients.[4,11]

Although congenital pseudoaneurysm is a rare finding on fetal echocardiography, it is important to consider this diagnosis when presented with an atrial mass on prenatal echocardiography, as this will help direct counseling for families prenatally and guide postnatal management.

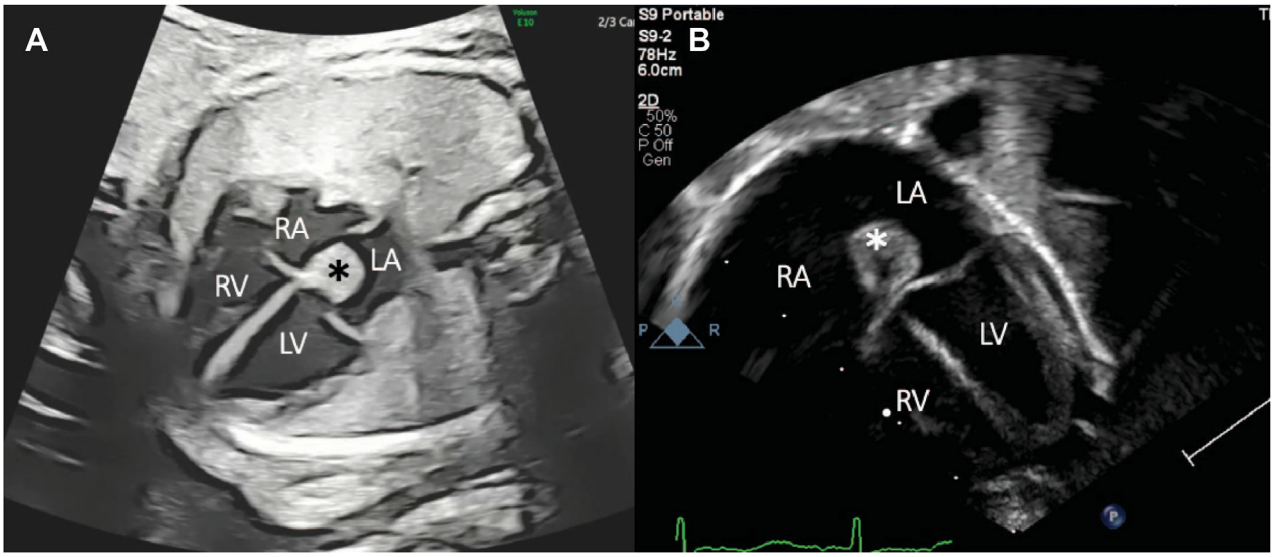

Figure 1 **(A)** Fetal two-dimensional echocardiographic image on initial study at 31-1/7 weeks' gestation in the four-chamber view, in systole, demonstrating a large echogenic mass (*asterisk*) in the anterior left atrium associated with the septum near the crux measuring 8.1 × 4.3 mm. **(B)** Postnatal two-dimensional transthoracic echocardiogram image on the first day of life in the apical four-chamber view, in systole, demonstrating location of the echogenic mass (*asterisk*) postnatally at the atrioventricular junction along the atrial septum, measuring 7.4 × 9.3 mm. *Asterisk* indicates P-MAIVF. *LA*, Left atrium; *LV*, left ventricle; *RA*, right atrium; *RV*, right ventricle.

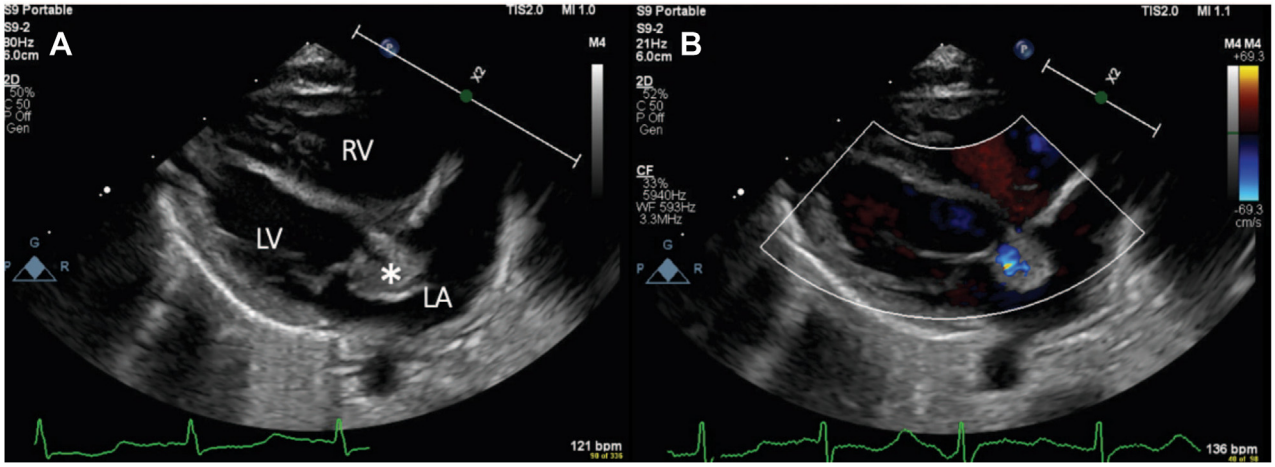

Figure 2 **(A)** Two-dimensional parasternal long-axis view, in systole, on postnatal transthoracic echocardiography on the first day of life showing the P-MAIVF (*asterisk*) originating between the aortic and mitral valves **(B)**, with color Doppler showing flow into the P-MAIVF in systole in the parasternal long axis view. *Asterisk* indicates P-MAIVF. *LA*, Left atrium; *LV*, left ventricle; *RV*, right ventricle.

CONCLUSION

P-MAIVF is an uncommon finding in the pediatric population, with only a few cases reported prenatally. This is an important diagnosis to consider in the setting of an atrial mass on fetal echocardiography and to make note of the specific echocardiographic findings characteristic of this diagnosis to guide prenatal counseling and postnatal management.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

CONSENT STATEMENT

Complete written informed consent was obtained from the patient (or appropriate parent, guardian, or power of attorney) for the publication of this study and accompanying images.

FUNDING STATEMENT

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DISCLOSURE STATEMENT

The authors report no conflict of interest.

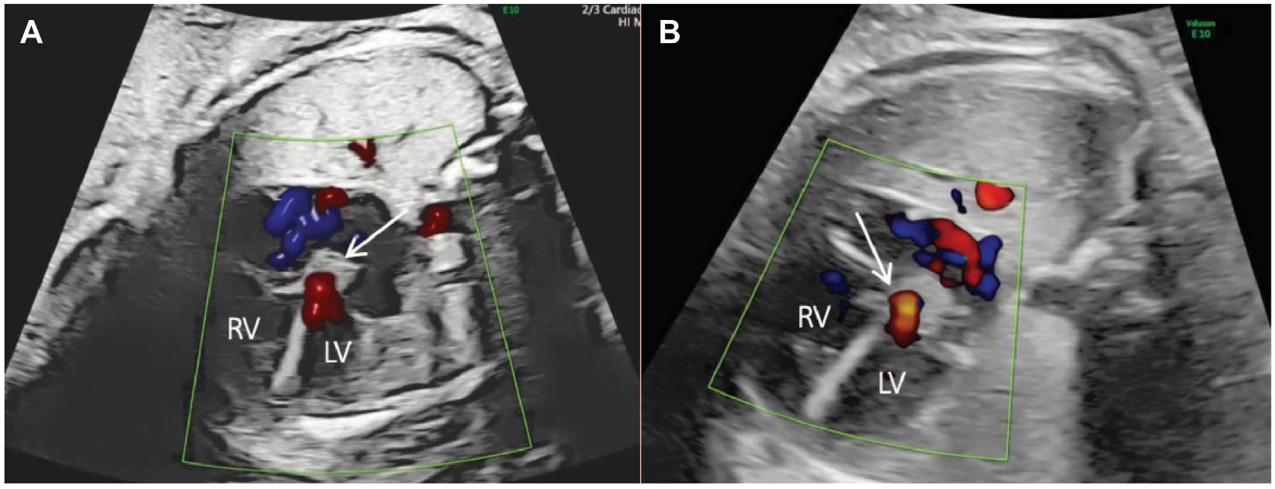

Figure 3 Fetal two-dimensional echocardiography with color Doppler in the four-chamber view demonstrating blood flow into the mass during systole (*arrow*) on initial study at 31 weeks' gestation **(A)** and follow-up study at 37 weeks' gestation, with blood flow into the mass during systole **(B)**. Blood flow into the mass was not initially noted on these studies, making this finding more consistent with a diagnosis of P-MAIVF. *LV*, Left ventricle; *RV*, right ventricle.

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2023.05.010.

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