Int Cardiovasc Res J.2018;**12**(3):121-123.icrj.65047

International Cardiovascular Research Journal
www.ircrj.com

Accessory "Pseudo" Left Atrium Confirmed by Cardiac Magnetic Resonance Imaging

Zahra Alizadeh Sani [1], Afsoon Fazlinezhad [2], Mohammad Vojdan-Parast [2], Shadi Sarafan-Sadeghi [3], Azin Seifi [3], Saeed Mirsadraee [4], Behshid Ghadrdoost [1], Mohaddeseh Behjati [5,*]

[1] Rajaie Cardiovascular Medical and Research Centre, Iran University of Medical Sciences, Tehran, IR Iran
[2] Cardiovascular Research Center, School of Medicine, Mashhad University of Medical Sciences, Mashhad, IR Iran
[3] Department of Medical Sciences, Mashhad Branch, Islamic Azad University, Mashhad, IR Iran
[4] University of Edinburgh, Little France Crescent, Edinburgh, United Kingdom
[5] Echocardiography Research Center, Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, IR Iran

ARTICLE INFO

Article Type:
Case Report

Article History:
Received: 12 Dec 2017
Revised: 14 Feb 2018
Accepted: 19 May 2018

Keywords:
Echocardiography
Magnetic Resonance Imaging

ABSTRACT

Accessory chamber is an extremely rare congenital abnormality, which is often detected as an incidental finding on echocardiography. We described a case of accessory pseudo Left Atrium (LA) firstly detected by echocardiography and confirmed by Cardiac Magnetic Resonance imaging (CMR). Case presentation- A 4-year-old girl referred to our center with recent fatigue and exertional dyspnea. A large accessory LA was clearly visible on transthoracic echocardiography. CMR demonstrated an isolated wide base round "auricle like" chamber with large pectinate muscles simulating bear's paw. The accessory LA's only connection was to the Left Ventricle (LV) through an orifice that functioned as a dysplastic valve at the level of Left Ventricular Outflow Tract (LVOT). This valve had severe regurgitation and stenosis with no papillary connection. Conclusion- This entity differs from cor-triatriatum of the LA because there is a superior-inferior septum between the two left-sided atria with no interatrial communication and no pulmonary venous drainage to the accessory chamber.

1. Introduction

Congenital Heart Disease (CHD) is common with an incidence of 0.8% of all live births (1). Cardiac Magnetic Resonance imaging (CMR) has an important role in anatomical and functional evaluation of CHD (2).

Accessory chamber is an extremely rare congenital abnormality that is often detected as an incidental finding on echocardiography, but patients may also present with symptoms. Herein, we describe a case of accessory pseudo Left Atrium (LA) firstly detected by echocardiography and confirmed by CMR, which has not been previously reported.

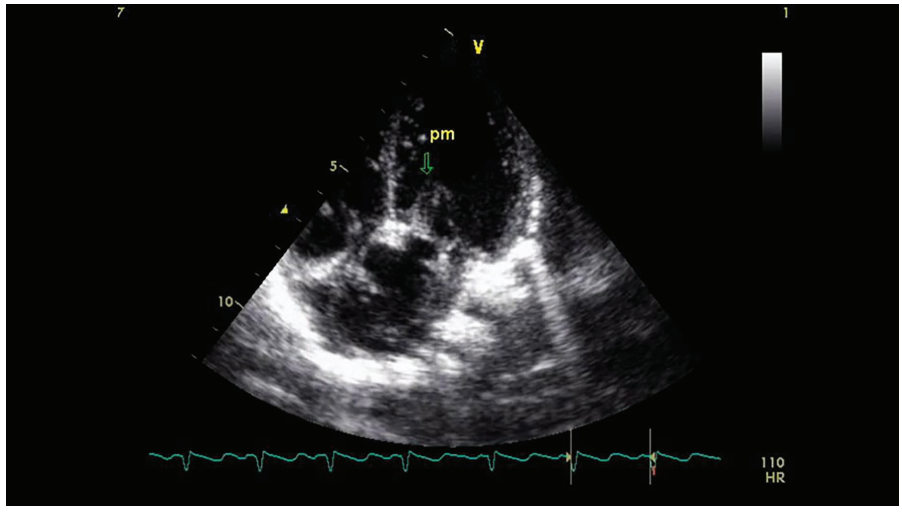
2. Case Presentation

A 4-year-old girl referred to our center with recent fatigue and exertional dyspnea. On physical examination, the patient was not cyanotic. Cardiac auscultation revealed a grade 3/6 holosystolic murmur accompanied with a grade 2/6 diastolic murmur on the left lower sternal border. Her blood pressure was 95/60 mmHg, heart rate: 102 beats/min, and the peripheral oxygen saturation was 94% at room air. A large accessory LA was clearly visible on Transthoracic Echocardiography (TTE) in the off axis Parasternal Long Axis (PLAX) and four chamber views (Figure 1). The accessory chamber had a round shape, measured 5.5 × 7.5 cm, and was located between the two atria. It communicated with the Left Ventricle (LV) via a restricted fixed orifice 5 mm in diameter (mean pressure gradient = 15 mmHg) with severe systolic regurgitation. Other TTE findings included left ventricular ejection fraction 55%, mild eccentric mitral regurgitation, normal aortic valve, mild Right Ventricle (RV) enlargement, moderate tricuspid regurgitation, mild pulmonary hypertension (pulmonary artery pressure = 45 mmHg), and a mildly dilated main pulmonary artery (Figure 1).

CMR imaging was performed for further evaluation using a 1.5 Tesla scanner (Avanto, Siemens, Erlangen) with a 6-element phased array receiver coil. CMR

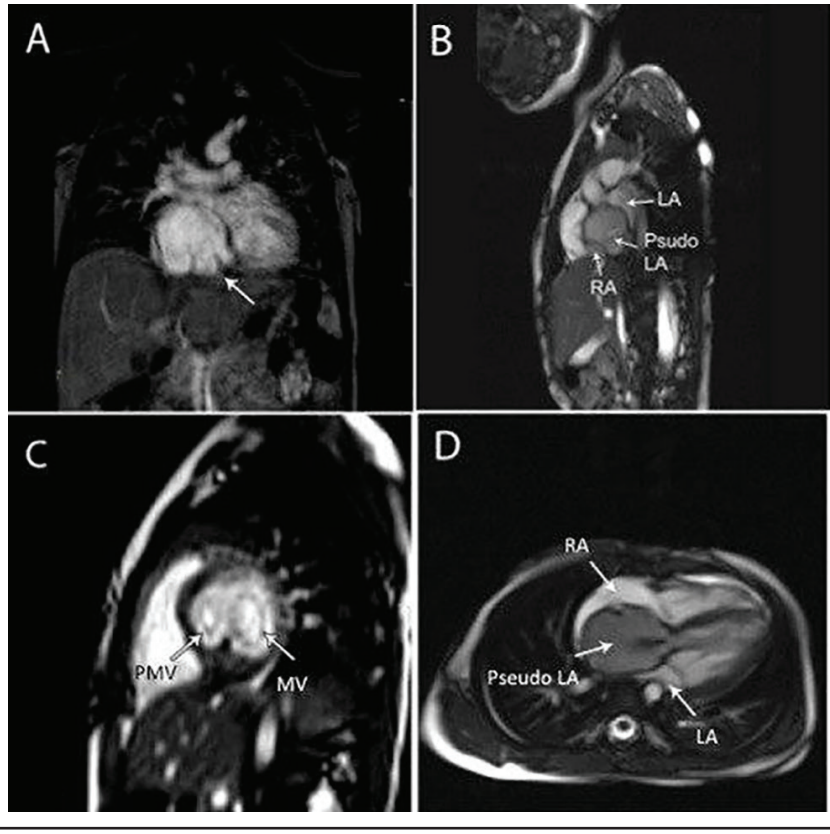
*Corresponding author: Mohaddeseh Behjati, Echocardiography Research Center, Rajaie Cardiovascular Medical and Research Center, Mellat Park, Vali-E-Asr Avenue, Tehran 1996911151, Iran, Tel: +98-2123922390, Fax: +98-2122663217, E-mail: dr.mohaddesehbehjati@gmail.com.

Figure 1. Echocardiographic Evaluation of Pseudo Left Atrium



Apical 4-chamber view demonstrating large central pseudo-atrium having relations with both atria.

Figure 2. CMR Images of the Heart (A: MR Angiography in Coronal Plain Showing a Large Lobulated Chamber (Arrow) with Pectinate-Like Muscle in Its Inferopostromedial Wall (Bear's Paw Appearance), B: Short Axis SSFP View in Basal Atrial Level Demonstrating Three Atria. Middle Chamber is the "Pseudo" Atrium Located between the Two True Atria., C: Short Axis SSFP Image Obtained during Diastole at the Level of Mitral Valve. Arrows Show Two Valve Orifices; the Right Arrow Shows Native AV Valve and the Left One Shows Dysplastic AV Valve, D: Four Chamber SSFP View Showing Two Left Atria with Significant Regurgitation Jet through the Dysplastic Valve)



Abbreviations: SSFP, steady-state free precession; AV, atrioventricular; PMV, pseudo-mitral valve; MV, mitral valve; RA, right atrium; LA, left atrium; LV, left ventricle; RV, right ventricle

demonstrated an isolated wide base round "auricle like" chamber with large pectinated muscles simulating bear's paw (Figure 2A). The chamber was located inferomedial to the LA (Figure 2B). All pulmonary veins drained into the LA properly. The accessory LA's only connection was to the LV through an orifice that functioned as a dysplastic valve at the level of Left Ventricular Outflow Tract (LVOT) (Figure 2C; Additional file 1). This valve had severe regurgitation and stenosis (orifice area = 25 mm2) with no papillary connection (Figure 2D). No concomitant cardiovascular anomalies were identified.

3. Discussion

An accessory left atrial chamber connected to the LV has been previously described. The cause was reported to be left atrial dissection, most commonly iatrogenic (e.g. post mitral or aortic valve surgery) (3) and rarely acquired as the result of infective endocarditis (4), myocardial infarction (5), or blunt chest injury (6). Spontaneous dissection as a result of a gap from the annular area to interatrial septum creating a new chamber has also been described previously. In some instances, mitral annular calcification was thought to be the origin of the dissection in the left atrial wall (7). Our patient had neither a history of previous surgery or trauma nor a potential cause for spontaneous dissection. Therefore, it was assumed that the accessory chamber might have been a congenital anomaly rather than an acquired abnormality.

Our patient was symptomatic with echocardiographic evidence of pulmonary hypertension. It has been reported that an increase in the left atrial pressure caused by the accessory atrium resulted in pulmonary hypertension and subsequently right ventricular dilatation with tricuspid regurgitation (4). Thus, we referred our patient for consideration of surgical intervention. The parents of the child, however, chose not to pursue surgical repair at this stage. Medical therapy with an Angiotensin-Converting Enzyme (ACE) inhibitor (Captopril) was then commenced.

3.1. Conclusion

This case was unique and, to the best of our knowledge, was the first CMR report of an accessory LA. This entity differs from cor-triatriatum of the LA because there is a superior-inferior septum between the two left-sided atria with no interatrial communication and no pulmonary venous drainage to the accessory chamber. Cardiologists have to be aware of such extremely rare anomalies that can potentially be associated with functional or thromboembolic complications.

Acknowledgements

There is no acknowledgement.

Authors' Contribution

Echocardiographic assessment and interpretation: Mohaddeseh Behjati, Afsoon Fazlinejad; Cardiac MRI assessment and interpretation: Zahra Alizadeh sani, Mohammad vojdan-parast, Shadi Sarafan-sadeghi; Manuscript writing: Behshid Ghadrdoost, Saeed Mirsadraee; Final approval of manuscript: Mohaddeseh Behjati, Zahra Alizadeh sani, Azin Seifi

Financial disclosure

There was no financial disclosure.

Funding/Support

There was no funding/support.

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