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Case Report



A rare case of double chamber left ventricle misdiagnosed as a pseudo-aneurism in cardiac magnetic resonance imaging: Case report

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Summary

A double chambered left ventricle (DCLV) is an extremely rare congenital malformation. In this anomaly left ventricle (LV) is divided into two sub-sections by a muscle bundle or a septum: main left ventricular chamber (MLVC) and accessory chamber. The reported prevalence of DCLV is 0.04%-0.42%; however, the precise prevalence is not determined. In this case, we are reporting a 54 years old man who was referred to cardiologist with a complaint of chest pain. He had a history of systemic hypertension. With regard to echocardiographic findings, the patient was diagnosed to have a left ventricular aneurism or DCLV. For further evaluation, cardiac magnetic resonance imaging (CMRI) was recommended. The results of CMRI revealed pseudo-aneurism with laminated thrombus. Nonetheless, through cardiac surgery DCLV with thick bundles was diagnosed. DCLV is rarely detected in adult patients since, in case of presence, it is mainly diagnosed during infancy or early childhood. Echocardiography is the initial imaging tool for diagnosis of DCLV; however, CMRI improves the chance of accurate diagnosis of the condition by providing a more detailed view. **Keywords:** Double chamber left ventricle, Congenital heart disease, Echocardiography, Cardiac magnetic resonance imaging, Pseudo-aneurism

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Introduction

Outpouching of left ventricle (LV) is a rare condition with variable etiologies including congenital heart problems, complications of coronary artery disease and myocardial infarction (aneurisms and pseudo-aneurysms), LV false tendon, LV fibrosis and diverticula. Considering that determining the etiology is clinically challenging, imaging evaluations play a fundamental role in management of these cases. LV outpouching or subdivision is rare in comparison to right ventricle. The divided chambers are usually separated by muscle bands or anomalous septum.

Nowadays, new imaging modalities such as echocardiography, contrast enhanced echocardiography, 3-dimensional echocardiography and cardiac magnetic resonance imaging (CMRI) are useful in thorough assessment of structural complications. Through CMRI, it is also possible to observe ventricular contractility, inflammation and fibrosis.¹⁻⁵

Pathogenesis in congenital forms of double chambered left ventricle (DCLV) may be associated with hypoplasia of regional myocardial intra-trabecular sinusoid, intra myocardial aneurysm formation during myocardial embryogenesis, 6 incomplete ventricular trabecular regression or a variant of non-compaction LV. 7

Like other myocardial abnormalities such as hypertrophic or dilated cardiomyopathies, genetic causes may be involved.8

Other poorly defined terms exist as well in available literature including LV diverticulum, LV aneurysm, LV cysts and accessory LV chamber. The most important differential diagnoses of DCLV are LV diverticulum and LV aneurysm. LV diverticulum is distinguished from DCLV by its morphology; as it has a narrow neck connecting to LV cavity. During echocardiography, presence of subdivision of LV to two distinct contracting chambers could be a sign of DCLV; though, the accessory chamber has not necessarily contractile function in all the patients with DCLV.

LV true aneurysm is a saccular protrusion of LV wall with a wide neck and scarred /fibrotic tissue happening due to mechanical weakness and contains all three cardiac layers. Aneurysms appear in echocardiography as an akinetic or dyskinetic segment in LV. ¹⁰ LV pseudo-aneurysm does not contain all the three layers of myocardium and has a high probability of rupture. Echocardiographic appearance of a pseudo-aneurysm is a dyskinetic segment. ²

Herewith, we describe a case of DCLV, with an emphasis on the importance of imaging in addition to echocardiography.

Case Presentation

A 54 years old man was referred to cardiologist with a complaint of recent non-anginal chest pain and





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palpitation. His past medical history revealed stage 1 systemic hypertension which was controlled by a daily dose of 25 mg losartan and a hospital admission due to anteroseptal myocardial infarction. Findings of physical examination were as: blood pressure=110/80 mm Hg, heart rate=75 bpm, muffled S1, normal P2 and no audible cardiac murmur. His electrocardiograph revealed incomplete left bundle branch block.

Echocardiographic evaluation unveiled normal LV size with global ejection fraction of 40% and large apical outpouching in size of 53 mm and a 25 mm neck. Color flow Doppler mode revealed bidirectional flow and mildly dilated ascending aorta. (Figures 1-3 demonstrate the Echocardiography views obtained from the patient.)

Echocardiologist recommended CMRI regarding that the accessory chamber was free of clot and had occasional contractions. CMRI was performed with 1.5 tesla, 18 canal Avanto Siemens scanner. In the results, LV apical pseudoaneurysm with laminated thrombus was reported. In coronary angiography a non-significant lesion in distal part left ascending artery was present.

Concerning the diagnosis of apical pseudo-aneurysm, the patient underwent surgery and through the operation thick fibrotic mid LV ring with aneurysmal apex were found.

Patient post-operative course was uneventful and follow up echocardiography showed an ejection fraction of 40%.

Discussion

DCLV is characterized by the sub-division of the LV chamber into two chambers by abnormal muscular tissue. It is differentiated from LV aneurysm and pseudoaneurysm by visualization of contractile function during systole in both chamber and also slight expansion due to increased pressure during systole.^{11,12}

In contrast to double chamber right ventricle, which is associated with septal defect, tetralogy of Fallot and/or transposition of great arteries, DCLV is commonly silent. In DCLV, there is a low pressure gradient between two chambers because of parallel position of main and accessory chambers. From etiologic point of view, DCLV is less known; but it is thought to be congenital and non-progressive. ^{13,14}

CMRI with normal delayed enhancement images is suggestive for ruling out coronary artery diseases and fibrosis.

Usually, DCLV is an incidental finding in the course of cardiovascular imaging. It is a rare condition with no definite available data regarding the outcome, prognosis and risk of embolism; nevertheless, it is generally believed that thromboembolic risk is low and treatment is guided by the presence of associated abnormalities.^{3,15,16}

We believe that the misdiagnosis of DCLV during CMRI was probably due to a poor thoracic window that

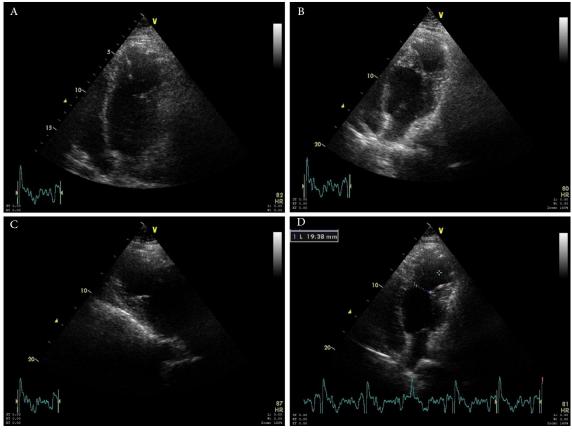


Figure 1. Apical chamber views. A, B, D represented an apical four chamber views and figure C revealed apical long axis view.

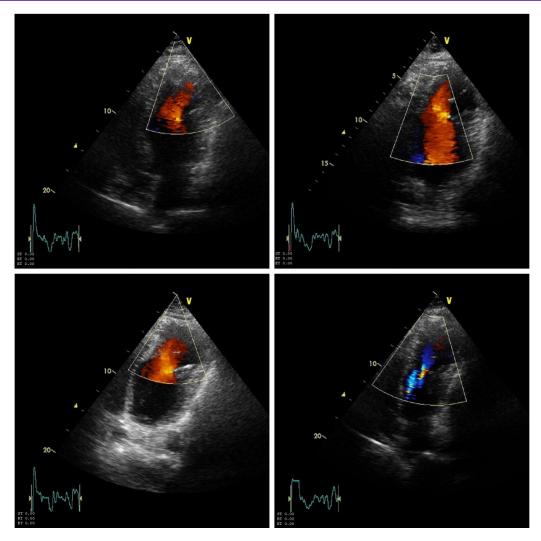


Figure 2. Color doppler flow imaging of left ventricular of the same patient.

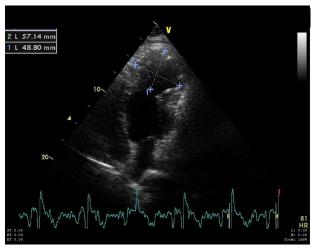


Figure 3. Left ventricular apical aneurysm size in four chamber views.

did not provide proper diagnostic-quality images of this condition.

Conclusion

Double Chamber Left Ventricle is a rare cardiac

malformation which is characterized by LV subdivision to main and accessory LV cavities. Usually being diagnosed during neonatal or pediatric age, it rarely remains asymptomatic until adulthood as in our case. Echocardiography can aid the detection of DCLV; however, CMRI allows a better evaluation and tissue characterization. In our case the coarse bundles inside the accessory chamber were confused with laying thrombosis in CMRI. With respect to the rarity of this anomaly, there are few evidence-based treatment approaches and the prognosis is poorly determined.

Authors' Contribution

Conceptualization: Rezvaniyeh Salehi, Rezayat Parvizi **Data curation:** Ramin Foroughi Asl, Mahsa Zehisaadat.

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Competing Interests

The authors declare no potential conflict o9f interest with respect to the research, authorship, and/or publication of this article.

Ethical Approval

Written informed consent was obtained from the participant for anonymized patient information to be published in this article.

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