Case Report

Myocardial Cleft, Crypt, Diverticulum, or Aneurysm? Does It Really Matter?

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ABSTRACT

Myocardial clefts are congenital abnormalities related to myocardial fiber or fascicle disarray that have been described in healthy volunteers as well as in the setting of hypertrophic cardiomyopathy. A cleft or crypt can be described as a discrete, approximately "V" shaped fissure extending into but confined by the myocardium, with a tendency to narrow or occlude in systole without local hypokinesia or dyskinesia. While little is known about the clinical significance of this entity, this report elaborates on the confounding terminology and differential diagnosis of this condition.

Introduction

Myocardial clefts are congenital abnormalities related to myocardial fiber or fascicle disarray, initially described in postmortem hearts in patients with hypertrophic cardiomyopathy.[1] More recently, cardiac magnetic resonance imaging (cMRI) has enabled in vivo detection of clefts in carriers of hypertrophic cardiomyopathy mutations[2] and healthy volunteers.[3] A cleft can be described as a discrete, approximately "V" shaped extension of blood signal penetrating >50% of the thickness of the adjoining compact myocardium in the long axis views, the cleft tending to narrow or occlude in systole, without local hypokinesia or dyskinesia.[3] We present a case of congenital myocardial cleft and discuss the diagnosis, confounding terminology, and significance of this entity.

Case Report

A 53-year-old male presented to the emergency room with progressive weakness and numbness of the right upper extremity for 4 d. He denied any chest pain or dyspnea. Patient admitted to noncompliance with antihypertensive medications, chronic 15 pack a year smoking history, and active cocaine abuse. He was afebrile with a heart rate of 98 bpm and a blood pressure of 116/89 mm Hg. His oxygen saturation was normal. There was no jugular venous distension. Cardiac exam revealed a dyskinetic point of maximal impulse, prominent left ventricular heave, and a regular rhythm. No murmurs or gallops were heard. Lungs were clear to auscultation. Moderate weakness of the right hand was also noted.

The electrocardiogram showed normal sinus rhythm, left axis deviation, ST-elevations in I, aVL, V_2 to V_6, and Q-waves in II, III, and aVF (Figure 1). Troponin I was >22 ng/ml (normal <0.4 ng/ml). CT of the head revealed multifocal infarcts in the internal capsule, lenticular nuclei, pons, and deep white matter of the left parietal lobe suggesting an embolic source.

Coronary angiography was notable for a total midleft anterior descending artery (LAD) occlusion. The left circumflex was a small vessel which was 100% occluded distally. The right coronary artery (RCA) had stenotic lesions (60–70%) in the proximal and distal segments. Percutaneous coronary intervention (PCI) and stenting of the LAD were performed. Left ventriculography revealed an akinetic anterior wall, an aneurysmal apex, and interestingly, a discrete, tubular, blind extension of the left ventricle (LV) cavity along the inferobasal wall, that opacified with contrast (Figure 1).

A transthoracic echocardiogram performed to further characterize this finding showed a small LV cavity, moderate concentric hypertrophy with septal and posterior wall thickness measurements of 1.6 and 1.5 cm, respectively. The mid to distal anterior wall was severely hypokinetic with apical dyskinesis. A discrete myocardial cleft was seen in the normally contracting mid-inferior wall. It measured approximately 2.1 × 0.4 cm in size and appeared to obliterate in systole (Figure 2A,C). Myocardial contrast echocardiography (MCE) with *Definity* (®Perflutren Lipid Microsphere, Lantheus Medical Imaging, N. Billerica, MA) showed preserved perfusion around this anomaly and significantly attenuated to absent myocardial perfusion in the anteroapical segments (Figure 2B,D). No intracardiac thrombus was identified; however, significant spontaneous echo contrast was visualized in the apical segment of the LV cavity. Systemic anticoagulation was initiated in view of the high likelihood of a cardiac source for the embolic stroke.

Clin. Cardiol. 32, 8, E48–E51 (2009)
Published online in Wiley InterScience. (www.interscience.wiley.com)
DOI:10.1002/clc.20466

Received: February 14, 2008
Accepted with revision: February 19, 2008

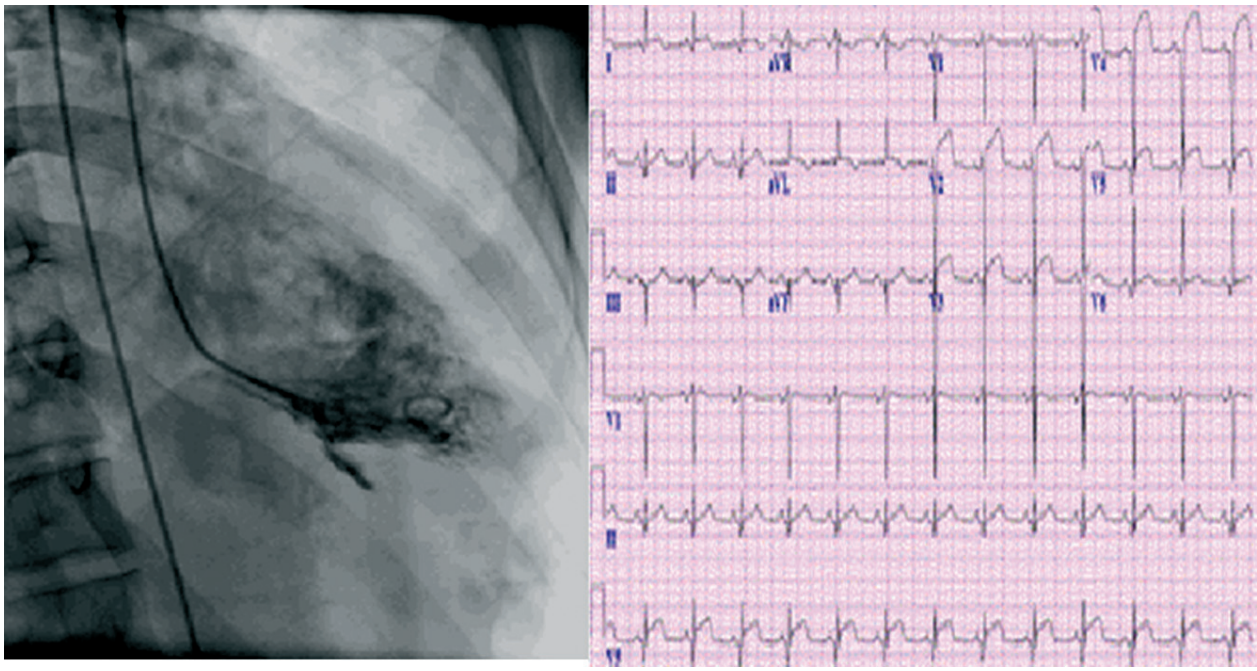

Figure 1. Ventriculogram (RAO projection)showing the contrast-filled myocardial cleft along the inferobasal wall. ECG with ST-segment elevation in the anterior precordial leads. *Abbreviations:* ECG = electrocardiogram; RAO = Right Anterior Oblique.

Discussion

Congenital myocardial clefts or fissures are commonly seen in the basal inferior wall of the left ventricle and the mid to apical segments of the interventricular septum. These regions are prone to developmental myocardial disarray, a histological abnormality of the crossing and interdigitation of the myocytes. In a study done by Germans et al., among 16 hypertrophic cardiomyopathy (HCM) mutation carriers, 13 of the subjects (81%) had evidence of clefts (called crypts in this study) in the basal and the mid inferoseptal myocardium although no evidence of hypertrophy was apparent on echocardiography.[2] These findings prompted the speculation that structural abnormalities like myocardial crypts might precede manifest hypertrophy in HCM mutation carriers. More recently, Johansson et al. reported 27 basal inferior and 24 septal myocardial clefts among the 399 cMRI cases studied retrospectively.[3] In this study, 13 of the 120 healthy volunteers (15.6%), 5 of the 91 HCM patients (5.5%), and 5 out of 44 hypertensive patients (11.4%) had clefts, suggesting that myocardial clefts are not limited to HCM genotypes. It is plausible that some of the clefts in subjects with overt HCM might have been compressed due to the development of hypertrophy, a finding not seen in gene mutation carriers. Clefts were seen in the long as well as the short axis planes, either single or in pairs with no apparent relationship to age, gender, LV function, or mass.

Myocardial clefts are congenital anomalies that probably have no prognostic significance. While clefts are diagnosed with cMRI relatively easily, echocardiographic visualization can be challenging, particularly if the cleft location does not coincide with standard acquisition planes. This case represents the first echocardiographically reported myocardial cleft. Proper alignment along the plane of the cleft enabled localization; further characterization was accomplished using contrast-enhanced harmonic imaging. The two-chamber apical view clearly profiled the cleft extending through most of the myocardium, contained only by a thin shell (~1 mm in thickness) of muscle that separated it from the right ventricle.

Myocardial clefts or crypts have to be differentiated from LV diverticulum, aborted myocardial rupture, pseudoaneurysm, and true LV aneurysm (Figure 3). Ventricular diverticula are congenital abnormalities of the myocardium, characterized by an outpouching of the myocardium in its entire thickness.[4] The incidence is reported to be 0.8% and while the majority are asymptomatic, systemic embolization, heart failure, valve regurgitation, and arrhythmias have been described.[5] Based on its location, a ventricular diverticulum can be classified as fibrous or muscular. Fibrous diverticula are subannular in location (submitral or subaortic), related to congenital annular weakness, and are not associated with cardiac and somatic anomalies. Muscular diverticula are usually apical, may occur in association with

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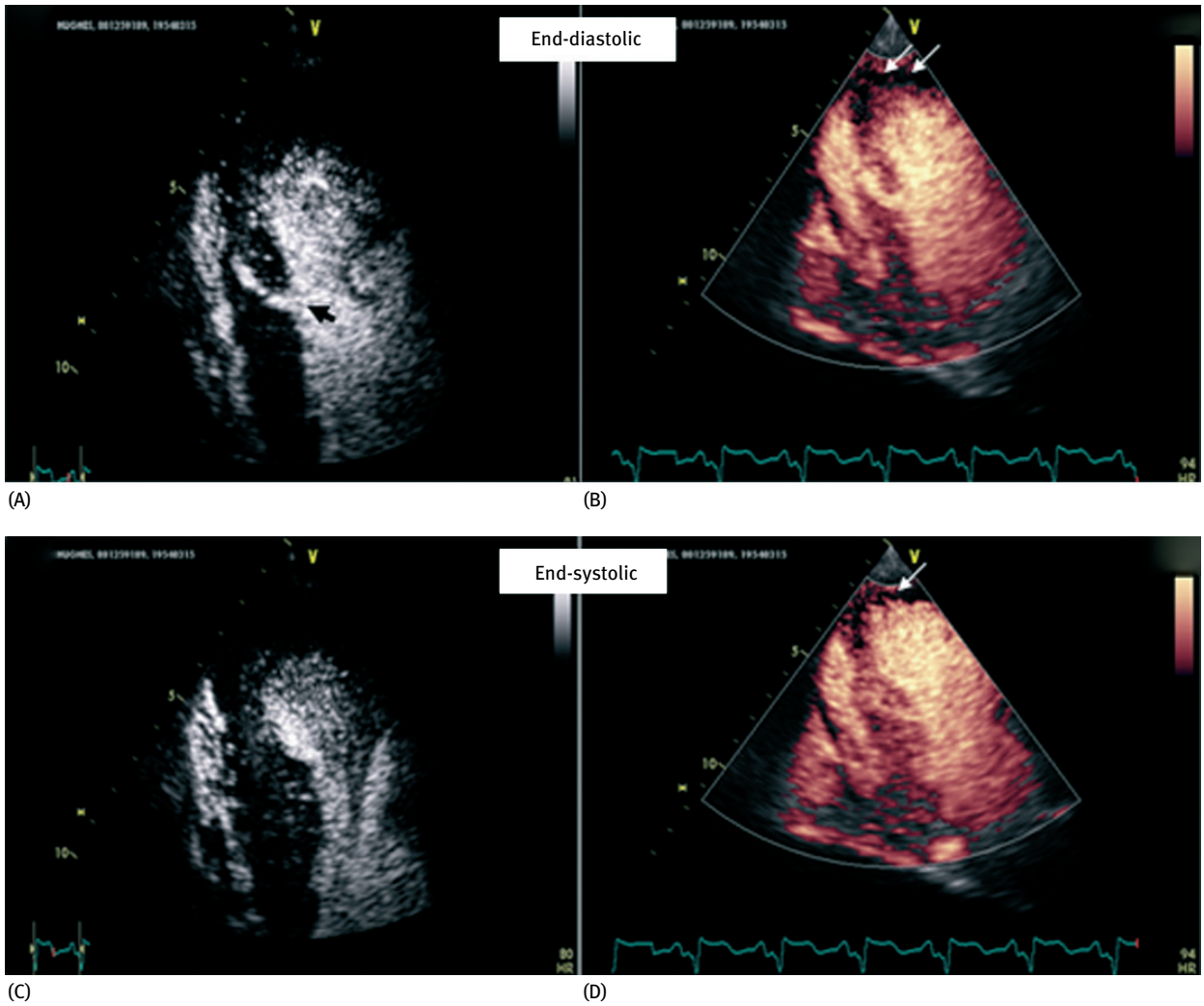

Figure 2. Off-axis, apical 2-chamber views in end-diastole (top panel) and end systole (bottom panel) delineating the myocardial cleft along the mid-inferior wall. Images A and C show contrast enhanced LV cavity with complete obliteration of cleft during systole. Myocardial contrast echocardiography (images B and D)depicting preserved perfusion around the cleft and grossly impaired perfusion at the apex (arrows). *Abbreviations:* LV = left ventricle.

cardiac and/or extracardiac defects such as Cantrell's syndrome and have all the cardiac layers. They arise due to defects in cardiac development or due to congenital myocardial thinning. Of note, both clefts and diverticula contract with systole, but diverticula typically are narrow mouthed with a wide outpouching, extending beyond the confines of the anatomic LV cavity and myocardial margin,[6] a morphological feature that helps differentiate them from clefts. Clefts, in contrast, are fissure-like protrusions confined to compacted myocardium that are contractile and may obliterate during systole. They also need to be differentiated from noncompacted myocardium, which is characterized by hypertrabeculated endocardium and a ratio of maximal thickness of the noncompacted to compacted layers of >2 (measured at end systole in a parasternal short axis view).[7]

Aneurysms of the ventricular wall that develop due to scarring are usually wide mouthed and thin walled and may exhibit characteristic paradoxical expansion or dyskinesis.[8] False aneurysms or pseudoaneurysms, on the other hand, are a form of myocardial rupture contained by the pericardium and typically result from infarction, trauma or tuberculosis. The cleft described in our patient was contained by a normally contracting well-perfused myocardium (confirmed on myocardial contrast echocardiography) supporting a congenital anomaly rather than an ischemic etiology.

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DOI:10.1002/clc.20466

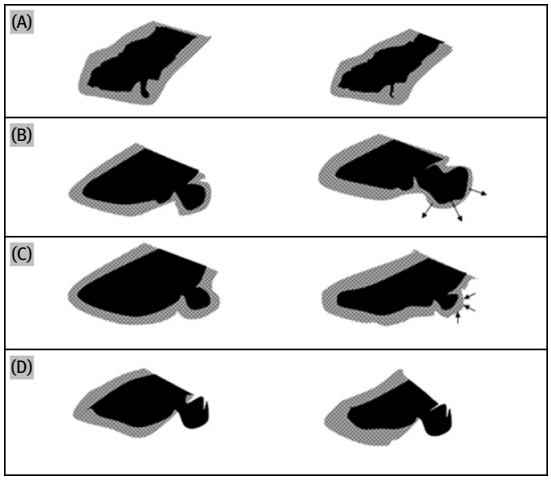

Figure 3. Schematic depicting myocardial cleft (A), ventricular aneurysm (B), ventricular diverticulum(C), and pseudoaneurysm (D) in diastole (left panel) and corresponding structural deformation during systole (right panel).

Conclusions

Myocardial clefts are distinct congenital anomalies that deserve recognition and have to be clinically differentiated for prognostic reasons from LV aneurysms and diverticula. Cardiac MRI and echocardiography represent the two best suited noninvasive imaging modalities for the accurate diagnosis of these entities.

References

1. Kuribayashi T, Roberts WC. Myocardial disarray at junction of ventricular septum and left and right ventricular free walls in hypertrophic cardiomyopathy. *Am J Cardiol*. 1992;70:1333–1340.
2. Germans T, Wilde AA, Dijkmans PA, et al. Structural abnormalities of the inferoseptal left ventricular wall detected by cardiac magnetic resonance imaging in carriers of hypertrophic cardiomyopathy mutations. *J Am Coll Cardiol*. 2006;48:2518–2523.
3. Johansson B, Maceira AM, Babu-Narayan SV, et al. Clefts can be seen in the basal inferior wall of the left ventricle and the interventricular septum in healthy volunteers as well as patients by cardiovascular magnetic resonance. *J Am Coll Cardiol*. 2007;50:1294–1295.
4. Tullu MS, Vaideeswar P, Deshmukh CT. Congenital left ventricular diverticula. *Int J Cardiol*. 2000;73:293–295.
5. Ohlow MA. Congenital left ventricular aneurysms and diverticula: definition, pathophysiology, clinical relevance and treatment. *Cardiology*. 2006;106:63–72.
6. Aquaro GD, Di Bella G, Strata E, et al. Cardiac magnetic resonance findings in isolated congenital left ventricular diverticuli. *Int J Cardiovasc Imaging*. 2007;23:43–47.
7. Weiford BC, Subbarao VD, Mulhern KM. Noncompaction of the ventricular myocardium. *Circulation*. 2004;109:2965–2971.
8. Krasemann T, Gehrmann J, Fenge H, et al. Ventricular aneurysm or diverticulum? clinical differential diagnosis. *Pediatr Cardiol*. 2001;22:409–411.

Published online in Wiley InterScience. (www.interscience.wiley.com)
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