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Case Report

A Case of Multiple Embolic Strokes Caused by a Congenital Left Ventricle Diverticulum Undetected on Echocardiography

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Abstract

Congenital Left Ventricular Diverticulum (CLVD) is a rare cardiac malformation caused by a localized protrusion of the left ventricular wall and usually diagnosed with routine echocardiography. CLVD is often associated with other cardiac and noncardiac abnormalities, but it can also occur alone. When echocardiography is non diagnostic, other noninvasive techniques such as cardiac CT (CCT) and Cardiac MR (CMR) can help to rule in/out the diagnosis by providing additional information on myocardial structure, morphology, and kinetics of both ventricles and of the diverticulum itself. We report the case of a patient who arrived in the Emergency Department for an acute cerebrovascular ischemic event with embolic pattern, who underwent non invasive diagnostic tests in order to identify the etiology.

Keywords: Cardiac malformation; Left ventricle wall abnormalities; Left ventricle diverticulum; Stroke; Echocardiography; Cardiac MR (CMR)

Abbreviation: CVLD: Congenital Left Ventricular Diverticulum; CCT: Cardiac CT; CMR: Cardiac Magnetic Resonance; CAD: Coronary Artery Disease; RV: Right Ventricle; LV: Left Ventricle; CLVC: Congenital Left Ventricle Cleft; LVA: Left Ventricle Aneurysm

Case Presentation

A 41-year-old female patient, with a history of recurrent stroke, arrived in the emergency department for the acute onset of hemilate deficiency; therefore, she was admitted to the Stroke Unit for therapy e diagnostic workup (Figure 1).

The first stroke episode dated to January 2019 during a hospitalization for measles in adulthood, the second one in July 2019. She reported a family history of ischemic heart disease (father), but she had no history of hypertension, diabetes mellitus, coronary artery disease (CAD) and cardiac arrhythmia. She reported easy fatigue and frequent headaches without aura. The physical examination was within normal limits and the electrocardiogram showed normal sinus rhythm. Bedside echocardiography was hampered by the high acoustic impedance of the thorax in forced supine decubitus, so that parasternal, apical and subcostal windows resulted inadequate for an optimal cardiac evaluation. Therefore, the patient underwent CMR due to a clinical suspicion of atrial and/or interventricular septal defect. CMR was performed with 1.5T scanner and with a standard protocol for cardiac morpho-functional evaluation. CMR ruled-out any inter-atrial or - ventricular septal defects, showing the presence of an abnormal long (3,5cm) saccular formation of the left apical interventricular septum, located medially the apex of the right ventricle (RV) (Figure 2), in absence of any left to right intracardiac shunt or intracavitary thrombotic formation. In addition, the described finding showed motion and systolic thickening synchronous with the remaining wall segments of the left ventricle (LV) placing the diagnostic suspicion of diverticulum. A collegial evaluation of the case in the context of the Heart-Team composed by cardiologists, cardiac surgeons, radiologists and neurologists excluded the surgical treatment, taking into account the small size of the diverticulum and given that the embolic episodes occurred in the absence of anticoagulant therapy. For these reasons an anticoagulant therapy was set.

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Discussion/Conclusion

Congenital left ventricular wall abnormalities detection has recently increased thanks to CMR or CCT examinations, both characterized by high sensibility to detect structural abnormalities sometimes undetected with echocardiography [1]. According to their morphology, motion during cardiac cycle, histological characteristics and outcomes, congenital left ventricular wall abnormalities can be divided into four major categories [2]

C**ongenital Left Ventricular Diverticulum (CLVD):** Saccular protrusion in continuity with the ventricular cavity and extending beyond the epicardial profile of the myocardial wall, with a narrow ventricular neck, presence of all the myocardial layers in its wall, and with normal and synchrony contraction with the remaining LV walls (Figure 3). CLVD is an uncommon condition in adults.

Congenital Left Ventricular Clefts (CLVC): "V-shaped" openings in the context of LV wall, and perpendicular to the long axis of the LV, penetrate more than 50% of the thickness of compact myocardium and obliterated during systole (Figure 4). This kind of wall abnormality is frequently referred to by other names including crypts, recesses, and fissures which are always depicted by LVC.

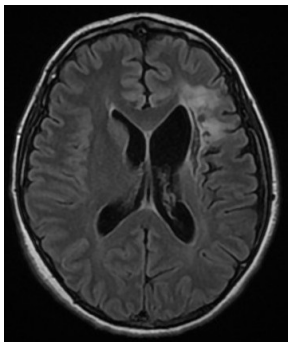

Figure 1: Axial FLAIR sequence of brain MRI shows the signs of previous ischemia in the left frontal and insular lobe with involvement of the homolateral corona radiata.

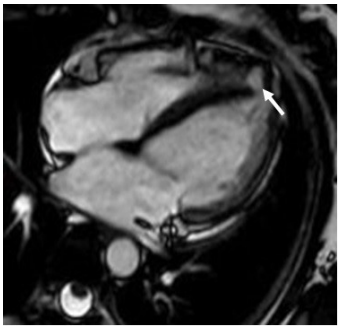

Figure 2: Horizontal long-axis cine bSSFP image in end-diastolic phase shows a congenital diverticular extroflection (length: 3.5cm-neck: 7mm, arrowhead) in the left ventricular septum in apical region, it is folded inferomedially and close to the right ventricular apex. Left ventricle non compaction myocardium is present.

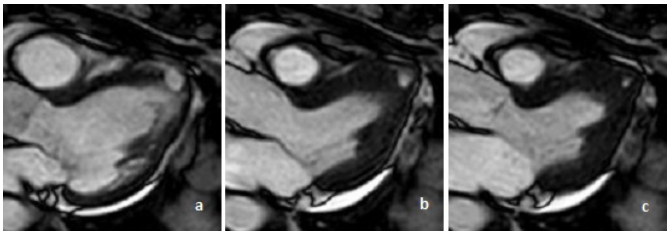

Figure 3: 3-chambers cine bSSFP images in end-diastolic (a), middle-systolic (b) and end-systolic (c) phases show sisto- diastolic parietal movement and thickening of the diverticulum, synchronous with other ventricular walls.

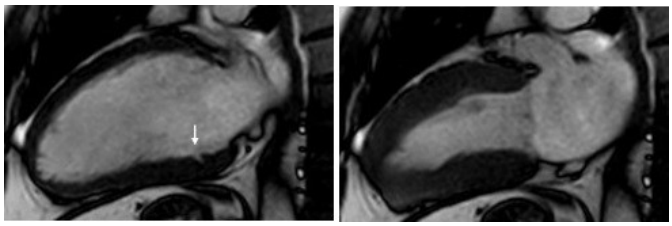

Figure 4: Vertical long-axis cine bSSFP (a) end-diastolic image shows V-shaped gap penetrating more than 50% of the thickness of compact myocardium, perpendicular to the long axis of the left ventricle; (b) the end-systolic image shows its obliteration during systole.

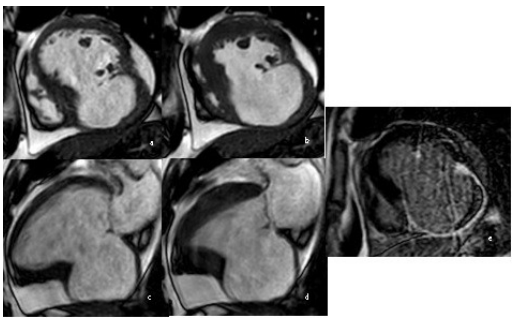

Figure 5: Short-axis (a, b) and vertical long-axis (c, d) cine bSSFP images in end-dyastolic (a,c) and end-systolic (b,d) phases show a large aneurysm in the mid-basal inferiorleft ventricular wall with a discinetic movement. Short-axis late gadolinium enhancement (e) show transmural LGE of the aneurysm's wall, with internal thrombus, which suggest an ischemic pattern.

Left Ventricular Aneurysm (LVA): Saccular out-pouching of the LV with a broad neck, thin fibrotic wall, typically akinetic or dyskinetic (Figure 5).

Pseudoaneurysm: Another left ventricular wall outpouching, arisen from a region of very thin or of absent myocardial wall as usually in case of recent and extended myocardial infarction and delimited by the pericardium [3].

CLVD is rare in adult population with an incidence around 0.4% in autopsy examination of adult patients with cardiac death, but the prevalence rises to 2.2% using newer imaging techniques [4]. Diverticulum may appear along the right and/or left ventrolateral borders of the endocardial tube in the fourth embryonic week following a developmental disturbance in the primordial paramidline mesoderm between the 14th and 18th day of the embryonic phase; for these reasons CLVD is often associated with other cardiac and midline thoraco-abdominal anomalies [1,5,6]. A partial halt in the growth of the fetal ventricle can explain the development of a congenital LVD and why the abnormality's position is commonly at the left ventricular apex, while non-apical locations are less frequent [7]. CLVD can be divided according to the structural characteristics into muscular or fibrous tissue, the latter with few or absent muscle fibers [7] and no volume change during cardiac cycle. The frequency of left ventricular apical wall abnormalities undetected by echocardiography is high because the cardiac apex is often difficult to evaluate with ultrasound. Congenital abnormalities of the left ventricular wall are often diagnosed incidentally by cardiac CT or cadiac MR performed for other clinical reasons. They are usually totally asymptomatic, especially when they are not associated with other congenital abnormalities, like in our patient, although CLVD and aneurysms have potentially harmful and even lethal complications as systemic embolization, ventricular arrhythmias, and sudden death due to ventricle rupture [1].

Despite the high frequency of absence of symptoms, there are cases in which the diagnostic delay may cause problems to patients such as in the illustrated case, in which the diagnostic delay contributed to the deterioration of the life's quality. Cardiac MR or CT provide accurate information about cardiac structures and allows easy recognition of congenital abnormalities of the LV wall that might otherwise be undetected.

This clinical case well shows, in our opinion, how important multimodality imaging is, in particular in patients suffering from multiple thromboembolic events of unknown cause. In these patients it is mandatory to avoid missing a diagnosis, even in the case of a rare condition such as CLVD.

In patients with a clinical suspicion of co-occurring events involving structural abnormalities of the LV wall,

any apparent structural normality on echocardiography should not make us feel confident and therefore prompt further higher-level imaging investigations, such as cardiac MR or CT.

In fact, other cases described in the literature have demonstrated that the existence of structural abnormalities of the LV wall can only be definitively ruled out through multimodal imaging, particularly regarding the apex [8]. Without a diagnosis of structural abnormality of the LV wall it is not possible to start any medical or surgical treatment to prevent the development of clinical manifestations related to it.

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