Case Report

A Rare Case of Coincidental LVNC and ALVC According to CMR

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ABSTRACT

Arrhythmogenic left ventricular cardiomyopathy (ALVC) is a rare inherited cardiomyopathy characterized by the progressive fibrofatty replacement of the left ventricular (LV) myocardium in combination with arrhythmias of LV origin. Herein, we present a case of the coincidence of ALVC and left ventricular non-compaction (LVNC) in a 34-year-old woman presenting with the chief complaint of palpitation. The patient had a history of an episode of severe common cold the previous year. Her mother was diagnosed with typical hypertrophic cardiomyopathy. The patient's preliminary echocardiography demonstrated evidence of LVNC. Subsequent evaluation using cardiac magnetic resonance (CMR) imaging showed a mildly enlarged LV without hypertrophy, along with noticeable myocardial trabeculation (Fig. 1) and a mildly decreased systolic function. Late gadolinium enhancement illustrated a fibrotic layer in the mid-anterior LV segment (Fig. 2). There was an elongated, thick stripe of subepicardial fat deposition at the base-to-mid–anteroseptal, anterior, and anterolateral LV segments (Fig. 3 & Fig. 4). Right ventricular apical trabeculation was prominent, with a regular ventricular function. The CMR tissue characterization criteria indicated the coincidence of ALVC and LVNC. Advances in noninvasive imaging modalities have significantly improved the delineation of the morphologic appearance of ventricles in different cardiomyopathies, facilitating identification and diagnosis.
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KEYWORDS: Arrhythmogenic left ventricle, Left ventricular non-compaction, Cardiac magnetic resonance imaging

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Arrhythmogenic left ventricular cardiomyopathy (ALVC) is a rare inherited cardiomyopathy characterized by the progressive fibrofatty replacement of the left ventricular (LV) myocardium in combination with arrhythmias of LV origin. [1] Herein, we present a case of the coincidence of ALVC and left ventricular non-compaction (LVNC).

Case Presentation

Our case describes a 34-year-old woman presenting with the chief complaint of palpitation. The patient had a history of an episode of severe common cold the previous year. Her mother was diagnosed with typical hypertrophic cardiomyopathy. The patient's preliminary echocardiography demonstrated evidence of LVNC. Subsequent evaluation with cardiac magnetic resonance (CMR) imaging revealed a mildly enlarged LV without hypertrophy, in addition to prominent myocardial trabeculation (Fig. 1) and a mildly reduced systolic function. There was an elongated, thick stripe of subepicardial fat deposition at the base-to-mid–anteroseptal, anterior, and anterolateral LV segments (Fig. 3 & Fig. 4). Right ventricular apical trabeculation was prominent with a regular ventricular function. Late gadolinium enhancement illustrated a fibrotic layer in the mid-anterior LV segment (Fig. 2). The CMR tissue characterization criteria indicated the coincidence of ALVC and LVNC.

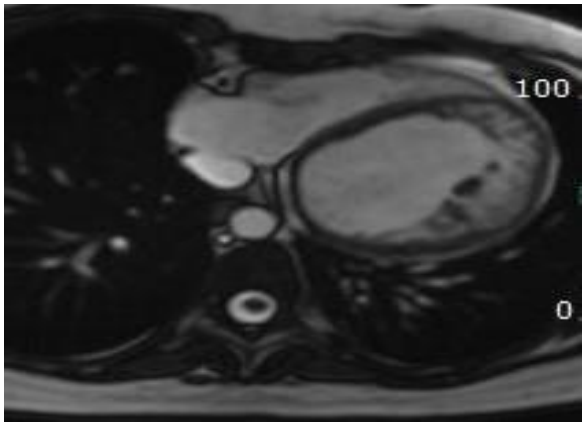

Figure 1: The cine steady-state free precession image shows a non-compacted left ventricular myocardium at the inferolateral wall.

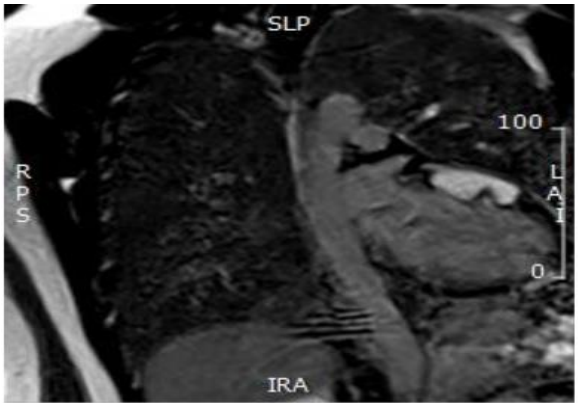

Figure 2: The late gadolinium enhancement sequence shows a thick intramural stripe of myocardial enhancement (bright signal) at the base-to-mid–anterior left ventricular segments.

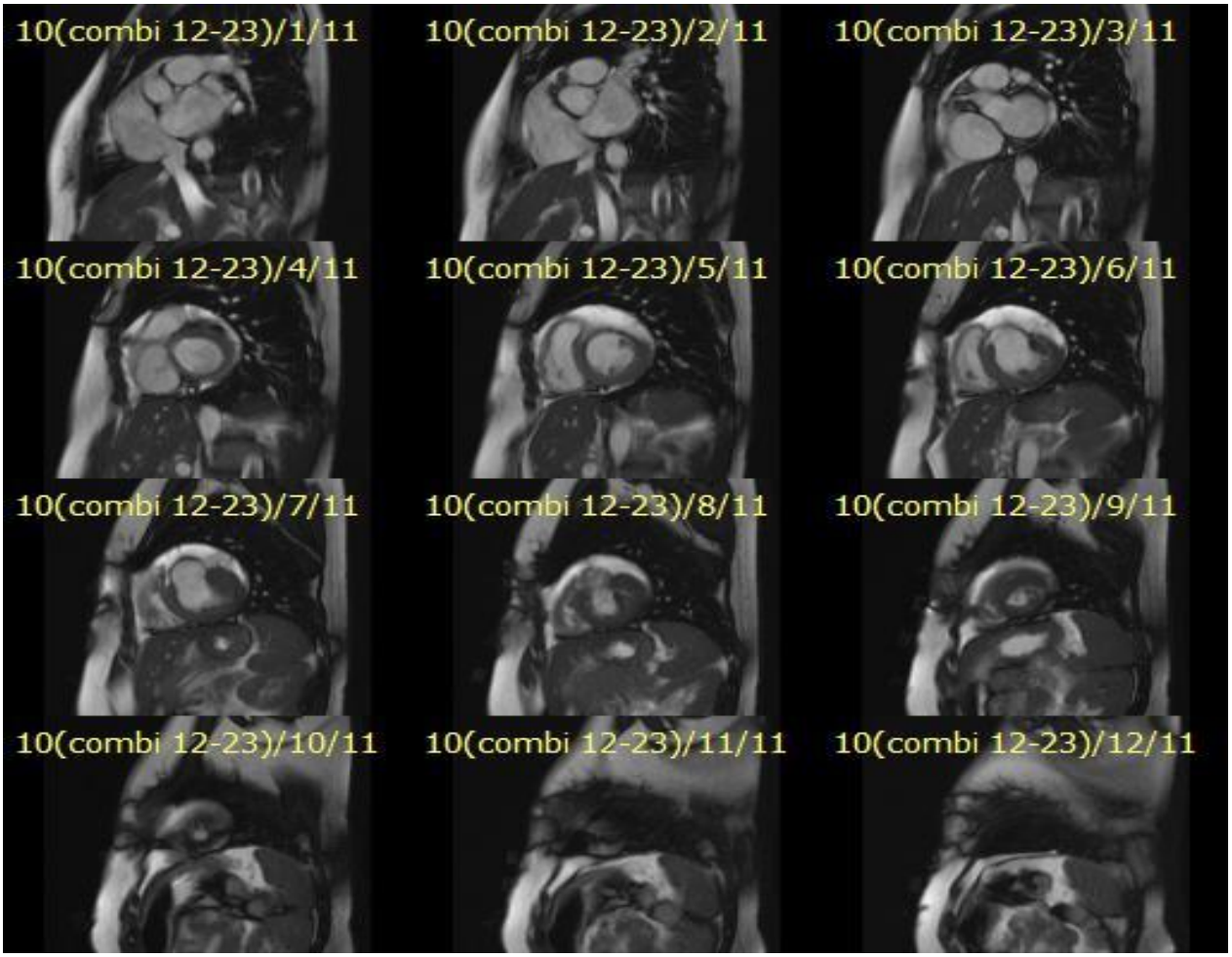

Figure 3: A stack of left ventricular short-axis steady-state free precession images demonstrates aneurysmal thinning and thick layers of subepicardial fat replacement at the base and mid-anterior left ventricular segments

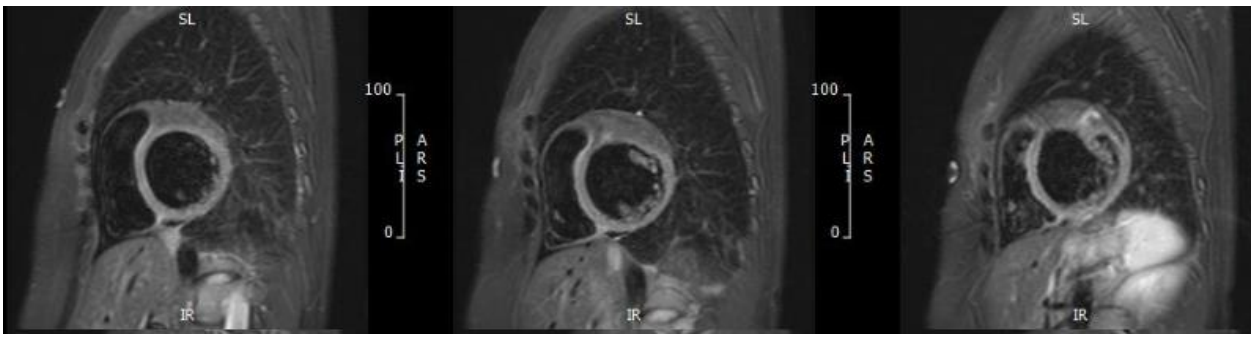

Figure 4: The turbo spin echo T1-weighted image with fat saturation sequences in the left ventricular short-axis plane reveals a thick stripe of subepicardial low-signal intensity at the anterior left ventricular wall.

DISCUSSION

ALVC is a rare congenital myopathy due to progressive myocardial fibrofatty deposition. [2] Although LV involvement in the setting of ALVC could be observed in advanced right ventricular dominant ALVC, left dominant type ALVC is very rare and lacks specific diagnostic criteria, making the diagnosis challenging. [3,4] LVNC is also a

very rare kind of congenital cardiomyopathy owing to impaired endomyocardial trabeculation, which carries a high risk of thromboembolism, arrhythmias, and ventricular dysfunction. [5] By now, the association of these 2 rare cardiomyopathies is rarely reported, which requires high physician vigilance. In this regard, advances in noninvasive imaging modalities have significantly enhanced the delineation of the morphologic appearance of ventricles in different kinds of cardiomyopathies, facilitating identification and diagnosis.

CONCLUSIONS

The co-occurrence of ALVC with LVNC is an extremely uncommon association. CMR using tissue characterization criteria is a very useful tool for the diagnosis of this rare association.

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