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Case Report

Base-to-apex gradient pattern of cardiac impairment identified on myocardial T1 mapping in cardiac amyloidosis

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ABSTRACT

Late gadolinium enhancement imaging by cardiac magnetic resonance imaging (CMR) is the most reliable method for identifying cardiac involvement in patients with amyloidosis, and myocardial T1 mapping is a novel CMR technique that enables the noninvasive detection and quantification of myocardial amyloid burden. Although, base-to-apex gradient patterns of impairment in patients with cardiac amyloidosis have been reported on myocardial strain analysis using echocardiography, we could not find any other reports to demonstrate that myocardial T1 mapping on CMR can clearly identify a base-to-apex gradient pattern of cardiac impairment in a patient with cardiac amyloidosis.

Introduction

Cardiac amyloidosis (CA) is considered a rare disease; however, recent research has revealed a considerable number of hidden CA cases, particularly in patients with heart failure having a preserved ejection fraction [1]. Cardiac involvement signifies a poor prognosis in patients with systemic amyloidosis. However, recent advancements in therapeutic interventions have contributed to an improved prognosis in these patients [2]. Thus, precise diagnosis and intervention with effective therapies are important. Advanced diagnostic imaging modalities, such as cardiac magnetic resonance imaging (CMR) facilitate a precise diagnosis of CA, allowing for the appropriate management of patients with CA [3].

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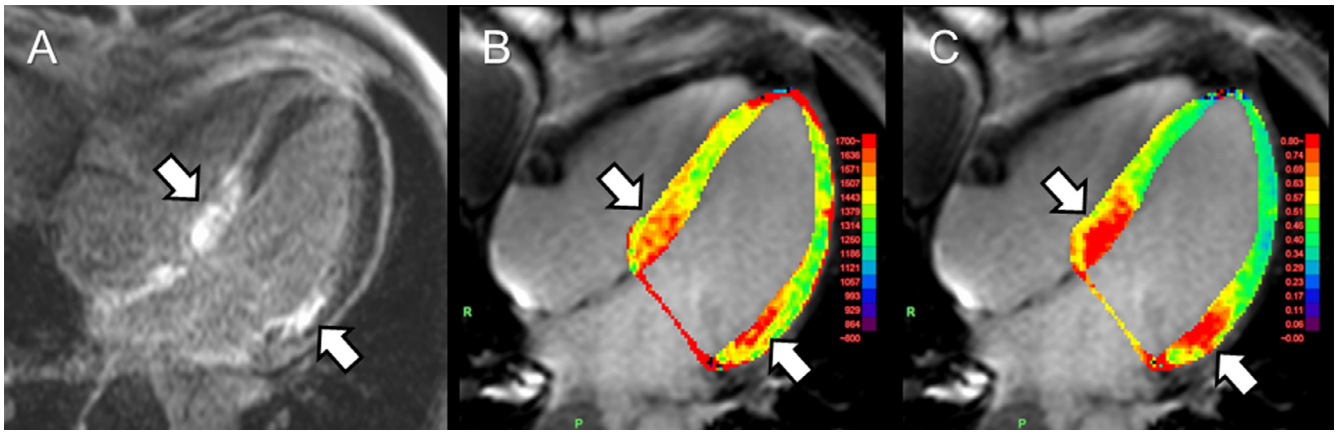

Fig. 1 – (A) Late gadolinium enhancement imaging revealed greater late enhancement at the base than at the apex, highlighting a base-to-apex gradient (arrows). Myocardial T1 mapping showed markedly elevated native T1 (B) and an increased extracellular volume fraction (C) at the base, as well as a base-to-apex gradient impairment pattern (arrows).

Case report

A 55-year-old man presented with proteinuria and lower extremity oedema. Transthoracic echocardiography demonstrated concentric left ventricular hypertrophy with a preserved ejection fraction of 66%. His plasma B-type natriuretic peptide level was elevated (79.7 pg/mL). CMR revealed greater late gadolinium enhancement (LGE) at the base than at the apex, highlighting a base-to-apex gradient (Fig. 1A). Myocardial T1 mapping showed markedly elevated native T1 and an increased extracellular volume fraction (ECV) at the base, as well as a base-to-apex gradient (Fig. 1B and C). The patient was eventually diagnosed with advanced systemic light-chain amyloidosis along with CA owing to underlying multiple myeloma.

Discussion

LGE imaging is the most reliable method for identifying cardiac involvement in patients with amyloidosis. The typical LGE pattern of CA is global subendocardial LGE with a dark blood pool [4]. However, recent studies have revealed a more variable LGE pattern in CA [5]. Some studies have demonstrated that a significant correlation of LGE with clinical, morphological, functional, and biochemical markers facilitates the evaluation of the prognosis of CA [6,7].

Myocardial T1 mapping (native T1 and ECV) is a novel CMR technique that enables the noninvasive detection and quantification of myocardial amyloid deposition and fibrosis. CA exhibits significantly higher native T1 and ECV values than those exhibited by other cardiac diseases, demonstrating a high diagnostic precision for the detection of CA [8]. Besides, native T1 and ECV values were elevated even in patients in whom conventional clinical testing and LGE imaging suggested no cardiac involvement, which emphasises the potential role of myocardial T1 mapping as an early diagnostic marker [9,10]. In addition, it helps to track various markers of disease activity, such as cardiac function and blood biomarkers, implying their correlation with the severity of CA [10,11]. Moreover, the assessment of myocardial T1 mapping aids in the risk-stratification of patients with CA [7,12], which may add incremental value over the existing clinical markers.

Our findings of a base-to-apex gradient pattern of impairment of native T1 and ECV values are in line with previous results showing that amyloid burden predominates at the base on myocardial strain analysis using two-dimensional speckle-tracking echocardiography [13] and cardiac uptake on bone scintigraphy with ^{99m}Tc-hydroxymethylene diphosphonate [14]; it is recognised as a clue to differentiate CA from other types of left ventricular hypertrophy. We could not find any other reports to demonstrate that myocardial T1 mapping on CMR can clearly identify a base-to-apex gradient pattern of cardiac impairment in a patient with CA.

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