

Short Report

Open Access, Volume 3

A unique pitfall of myocardial perfusion imaging in transthyretin amyloid cardiomyopathy

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Abstract

Myocardial Perfusion Imaging (MPI) is a non-invasive modality with proven diagnostic and prognostic utility in coronary artery disease. We present a case of Transthyretin Amyloid Cardiomyopathy (ATTR-CM) where MPI findings were vastly different from Transthoracic Echocardiogram (TTE) and Cardiac Magnetic Resonance Imaging (CMR). A 78-year-old gentleman presented with atypical chest pain to rule out ischemic heart disease. He underwent MPI scan, revealing a medium-sized fixed perfusion defect of moderate intensity in the inferior segments. Post-stress gated Left Ventricular Ejection Fraction (LVEF) and left ventricle End-Diastolic Volume (EDV) were estimated to be 33% and 205 ml. On the same day, TTE showed left ventricular hypertrophy and systolic anterior motion of the anterior mitral leaflet, with preserved LVEF of 60% and EDV of 114 ml. In view of the discrepancies between the two tests, the patient was sent for a CMR scan. Overall impression of CMR was in keeping with infiltrative cardiomyopathy. Urine and serum amyloid screen were unremarkable. Subsequently, Technetium TC 99 M Pyrophosphate scan was performed and was highly suggestive of ATTR-CM. Through this case presentation, we explore the pitfalls of myocardial perfusion imaging in patients with ATTR-CM and careful interpretation of results is needed in these selected group of patients.

Received: Jul 26, 2022

Accepted: Aug 19, 2022

Published: Aug 26, 2022

Archived: www.jcimcr.org

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DOI: www.doi.org/10.52768/2766-7820/2017

Keywords: Myocardial perfusion imaging; Transthyretin amyloid cardiomyopathy; Transthoracic echocardiogram; Cardiac magnetic resonance; Technetium tc 99m pyrophosphate scan.

Abbreviation: MPI: Myocardial Perfusion Imaging; ATTR-CM: Transthyretin amyloid cardiomyopathy; TTE: Transthoracic echocardiogram; CMR: Cardiac magnetic resonance imaging; LBBB: Left bundle branch block; LV: Left ventricular; LVEF: Left ventricular ejection fraction.

Introduction

Myocardial Perfusion Imaging (MPI) is a proven diagnostic and prognostic tool in coronary artery disease [1]. Besides information on coronary perfusion, Left Ventricular Ejection Fraction (LVEF) can be derived using Electrocardiogram (ECG) gating [2]. There are pitfalls in interpretation of MPI, requiring alternative imaging technique to corroborate abnormal observations [3]. We present a case of Transthyretin Amyloid Cardiomyopathy (ATTR-CM) where MPI findings were vastly different from that of Transthoracic Echocardiogram (TTE) and Cardiac Magnetic Resonance Imaging (CMR).

Case presentation

A 78-year-old gentleman with medical history of hypertension, hyperlipidemia, diabetes mellitus, osteoarthritis and glaucoma, presented with atypical chest pain. His ECG revealed Left Bundle Branch Block (LBBB) pattern and Left Ventricular (LV) hypertrophy by Cornell Criteria (Figure 1). No previous ECGs were available for comparison. Physical examination was unremarkable. Troponin I was mildly elevated up to 111 ng/L (Normal range 0-17.4 ng/L). His creatinine level was raised at 134 umol/L (baseline 60-107 umol/L). TTE showed normal Left Ventricular Ejection Fraction (LVEF) of 60% and confirmed marked concentric LV hypertrophy on TTE (Figure 2). Using modified Simpsons biplane method of disks, LV end-diastolic volume and LV end-systolic volume were 114 and 52.8 mls, respectively. On the same day, he underwent a dipyridamole stress MPI scan to exclude underlying ischemic heart disease. MPI revealed a medium-sized fixed perfusion defect of moderate intensity in the inferior segments (Figure 3). LVEF was 40% during rest and 33% post-stress. LV end-diastolic volume was 140 ml during rest, and 205 ml post-stress. Because of conflicting results in LVEF between TTE and MPI, he was sent for a CMR scan. CMR findings demonstrated LV hypertrophy and elevated LV mass index of 213 g/m². LVEF was 59% and LV EDV was 127 ml. There was extensive mid-wall fibrosis in the hypertrophied LV myocardium especially over the inferior and lateral walls, and patchy fibrosis of LV papillary muscles (Figure 4). These findings generally matched those of his TTE. Overall impression of CMR was in keeping with infiltrative cardiomyopathy. Urine and serum amyloid screen were unremarkable. Subsequently, Technetium TC 99 M Pyrophosphate scan was performed which was highly suggestive of ATTR-CM (Figure 5). Alternative tests to exclude ischemic heart disease were considered but deemed unsuitable. Age-related coronary calcifications will obscure underlying coronary arteries on computer tomography. Invasive coronary angiogram was associated with significant risk of contrast-induced nephropathy in the setting of renal impairment.

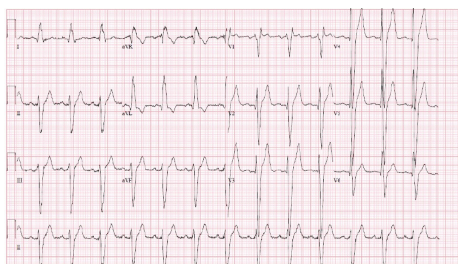


Figure 1: 12-lead electrocardiogram demonstrating new onset left bundle branch block and left ventricular hypertrophy by the Cornell Criteria.

Figure 2A, 2B: Transthoracic echocardiogram demonstrating left ventricular hypertrophy in 4-chamber view and parasternal long axis view.

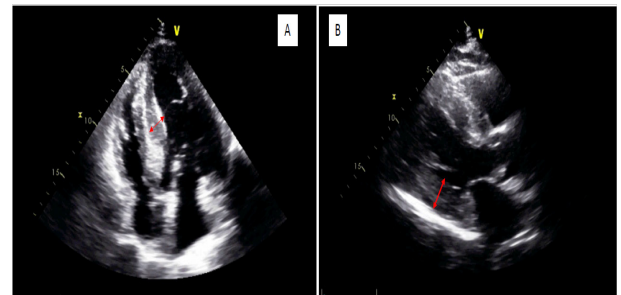


Figure 3A, 3B: Myocardial perfusion images demonstrating fixed perfusion defects in inferior segments.

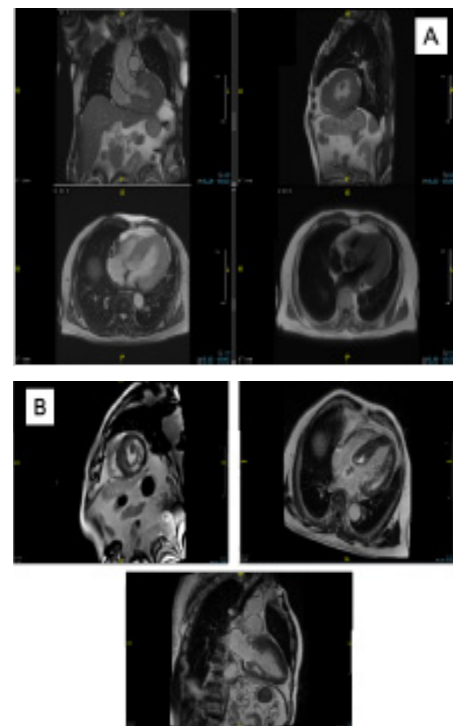


Figure 4: Cardiovascular magnetic resonance imaging demonstrating significant left ventricular hypertrophy in coronal, sagittal and axial planes.

Figure 4B: Cardiovascular magnetic resonance imaging demonstrating mid-wall fibrosis in left ventricular myocardium and patchy fibrosis of papillary muscles in short axis, 3-chamber and vertical long axis views.

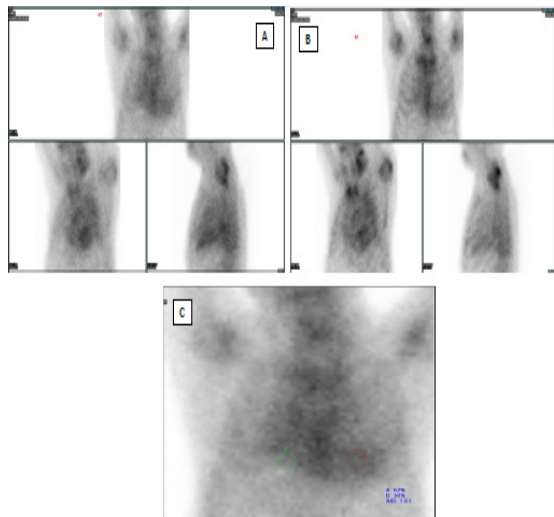


Figure 5: Technetium TC 99M Pyrophosphate scan planar images in the anterior, left anterior oblique and left lateral projections after intravenous injection of Tc-99m pyrophosphate.
 Figure 5A: Images taken at 60 minutes after injection.
 Figure 5B: Images taken 3 hours after injection.
 Figure 5C: Quantitative myocardial to contralateral lung ratio of uptake at 60 mins was 1.61 at 60 minutes.

Discussion

The above case highlights the potential setbacks in interpretation of MPI images. Perfusion defects on MPI typically indicate coronary ischemia, but could occasionally be due to other reasons such as apparent perfusion abnormalities from myocardial fibrosis or LBBB as demonstrated in our patient. Firstly, perfusion defects on MPI may be a result of reduced tracer uptake from myocardial fibrosis as seen in our patient with ATTR-CM. Apart from ATTR-CM, other non-ischemic conditions such as hypertrophic cardiomyopathy or other infiltrative cardiac diseases like sarcoidosis that causes myocardial fibrosis can also result in apparent perfusion defects in MPI. Secondly, presence of LBBB also induces artefactual reversible septal perfusion, precluding useful interpretation of MPI [4]. In infiltrative cardiac diseases such as ATTR-CM, patients may develop LBBB as a manifestation of conduction abnormalities, resulting in false positive MPI [5]. Although LV size and LVEF can be derived from gated MPI scan, it may be inaccurate in patients in certain scenarios. Infiltrative cardiac diseases such as ATTR-CM are often associated with extensive mid-wall fibrosis and left ventricular hypertrophy with subendocardial hypoperfusion [5,6]. The presence of myocardial fibrosis and reduced perfusion results in reduced tracer count, adding to the poor spatial and temporal resolution of MPI [7]. With poor LV border detection, LV size is overestimated and subsequent LVEF underestimated in MPI, as illustrated in our patient with ATTR-CM. Hence, in comparison to other modalities, MPI may not be the best modality in the assessment of both LV function and volumes in patients with infiltrative cardiac diseases and must be used cautiously.

Conclusion

MPI is useful in evaluation of coronary ischemia, LV size and LVEF. However, clinicians should be cautious and judicious in the interpretation of MPI images with respect to certain scenarios such as infiltrative cardiac disease, taking into account the potential pitfalls.

References

1. Klocke FJ, Baird MG, Lorell BH, Bateman TM, Messer JV, et al. ACC/AHA/ASNC Guidelines for the Clinical Use of Cardiac Radionuclide Imaging— Executive Summary: A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (ACC/AHA/ASNC Committee to Revise the 1995 Guidelines for the Clinical Use of Cardiac Radionuclide Imaging). *J Am Coll Cardiol.* 2003; 42: 1318–1333.
2. Abidov A, Germano G, Hachamovitch R, Slomka P, Berman DS. Gated SPECT in assessment of regional and global left ventricular function: An update. *J Nucl Cardiol.* 2013; 20: 1118–1143.
3. Burrell S, Mac Donald A. Artifacts and pitfalls in myocardial perfusion imaging. *J Nucl Med Technol.* 2006; 34: 193–211.
4. Koepfli P, Wyss CA, Gaemperli O, Siegrist PT, Klainguti M, Schepis T, et al. Left bundle branch block causes relative but not absolute septal underperfusion during exercise. *Eur Heart J.* 2009; 30: 2993–2999.
5. Shah KB, Inoue Y, Mehra MR. Amyloidosis and the Heart: A Comprehensive Review. *Arch Intern Med.* 2006; 166: 1805–1813.
6. Hashimura H, Ishibashi Ueda H, Yonemoto Y, Ohta Ogo K, Matsuyama TA, Ikeda Y, et al. Late gadolinium enhancement in cardiac amyloidosis: Attributable both to interstitial amyloid deposition and subendocardial fibrosis caused by ischemia. *Heart Vessels.* 2016; 31: 990–995.
7. Pai M, Yang YJ, Im KC, Hong IK, Yun SC, et al. Factors Affecting Accuracy of Ventricular Volume and Ejection Fraction Measured by Gated Tl-201 Myocardial Perfusion Single Photon Emission Computed Tomography. *Int J Cardiovasc Imaging.* 2006; 22: 671–681.