



Multimodality imaging of simultaneous occurrence of cardiac transthyretin amyloidosis and cardiac sarcoidosis

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Received Jul 21, 2022; accepted Jul 23, 2022
doi:10.1007/s12350-022-03083-5

INTRODUCTION

We illustrate the findings of multimodality imaging (i.e., DPD-scintigraphy, 18F-fluorodeoxyglucose (18F-FDG) positron emission tomography (PET), cardiac magnetic resonance (CMR)) in endomyocardial biopsy proofed cardiac sarcoidosis and cardiac transthyretin amyloidosis (ATTR).

CASE PRESENTATION

A 79-year-old female presented with fatigue, dyspnea, elevated NT-proBNP, and recurrent exudative pleural effusion of unknown origin without bacteria or neoplastic cells. Pulmonary embolism, pneumonia, and tumor were ruled out by computed tomography. Echocardiography showed slightly reduced left ventricular ejection fraction with diffuse hypokinesia and apical sparing. Light-chain amyloidosis was ruled out and DPD-scintigraphy showed cardiac ATTR. As diagnostic thoracoscopy revealed no malignancy, but non-necrotizing granulomas in the lung, a FDG-PET was performed, which suggested multiorgan sarcoidosis. CMR showed diffuse fibrosis and multifocal late

gadolinium enhancement (LGE) suggesting cardiac ATTR. However, the extensive LGE and multifocal inflammation would also be in accordance with cardiac sarcoidosis. Endomyocardial biopsy was performed, and histological analysis confirmed the diagnosis of both cardiac sarcoidosis and cardiac ATTR (differentiation revealed wild-type ATTR). Tafamidis and immunosuppressant therapy were initiated.

DISCUSSION

The main differentials according to the imaging findings were (a) FDG-PET uptake in LV-myocardium: cardiac ATTR mimicking cardiac sarcoidosis?¹ (b) DPD-scintigraphy uptake in LV-myocardium: cardiac sarcoidosis mimicking cardiac ATTR?² (c) Simultaneous presence of both entities, which, to our knowledge, has not yet been described in the literature. Multimodality imaging and endomyocardial biopsy could establish the diagnosis of the simultaneous occurrence of cardiac ATTR and cardiac involvement of systemic sarcoidosis and helped to guide optimal medical therapy of this patient (Figure 1).

Funding Dr. Gräni receives funding from the Swiss National Science foundation, InnoSuisse, Center for Artificial Intelligence Grant from the University of Bern and the GAMBIT foundation.

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J Nucl Cardiol

1071-3581/\$34.00

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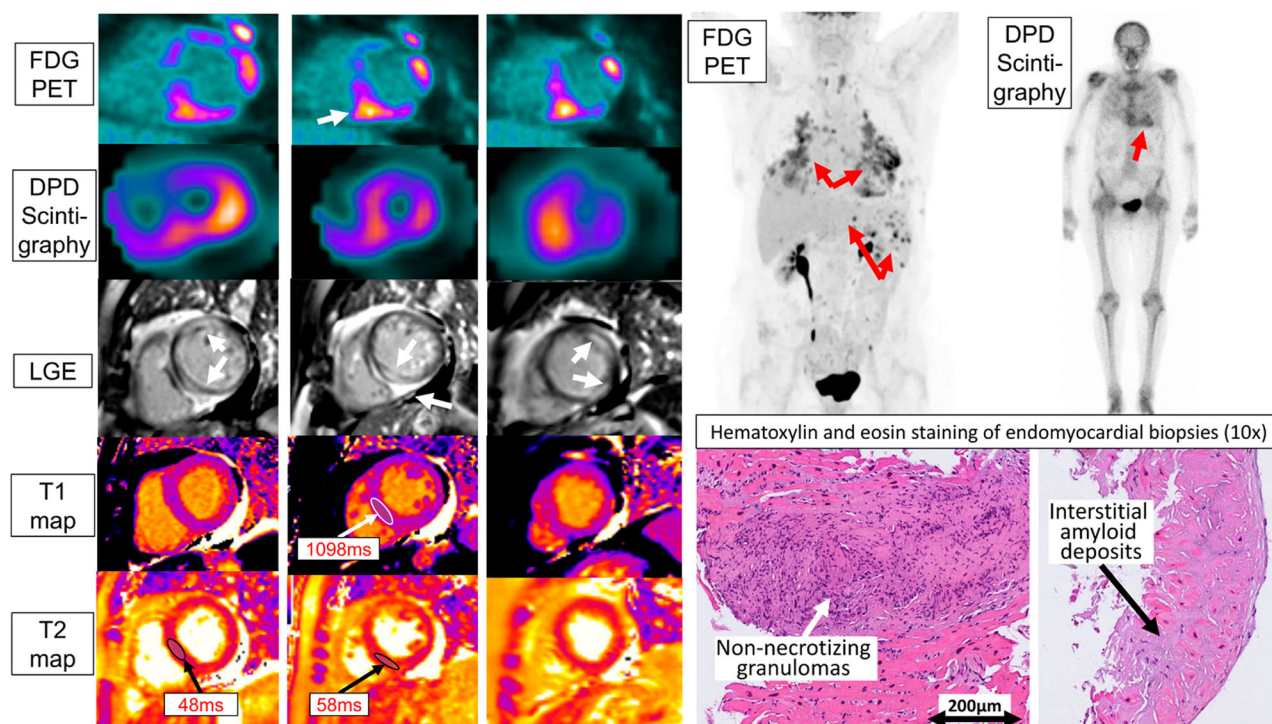


Figure 1. The cardiac short axis PET view on the upper left side shows multifocal FDG cardiac uptake. Multiorgan FDG uptake is depicted on the upper right side. DPD-scintigraphy showed Perugini grade 2 consistent with cardiac ATTR. LGE shows extensive scarring in in the endocardium, epicardium, midmyocardium, and diffuse fibrosis with elevated T1 mapping values and extracellular volume fraction (between 42 and 51%) were measured throughout the myocardium. T2 mapping shows multifocal edema in the myocardium. Hematoxylin, eosin, and congo red staining of endomyocardial biopsy confirmed simultaneous occurrence of cardiac sarcoidosis and cardiac amyloidosis (further differentiation revealed wild-type ATTR).

Disclosures

Dr. Caobelli receives grants and speaker honoraria from Siemens Healthineers and Bracco AG for matters not related to the present paper. All other authors have nothing to disclose.

Funding

Open access funding provided by University of Bern.

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References

1. Young KA, Lyle M, Rosenbaum AN, Chang IC, Lin G, Bois MC. (18)F-FDG/(13)N-ammonia cardiac PET findings in ATTR cardiac amyloidosis. *Nucl Cardiol*. 2022. <https://doi.org/10.1007/s12350-021-02886-2>.
2. El-Tallawi KC, Parikh R, Nabi F, Maclayton PI, Trachtenberg BH, Al-Mallah M. A positive Tc-99m PYP scan in a patient with cardiac sarcoidosis. *J Nucl Cardiol*. 2021;28:2390-4.

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