

Cardiac sarcoidosis detected with hybride imaging

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Abstract

Sarcoidosis is a systemic granulomatous disease of unknown cause. Approximately one fourth of all patients with sarcoidosis have clinical manifestations of cardiac involvement. Granulomas may involve the pericardium, myocardium and endocardium of both the ventricles, as well as the atria. Cardiac involvement is associated with poor prognosis and is definitely confirmed with endomyocardial biopsy. Given the invasiveness and risks of the procedure, other alternative diagnostic methods are used. In diagnosis and clinical assessment of disease activity broad spectrum of analysis is used: clinical examination, laboratory analysis (ACE, chitotriosidase), multi-detector computer tomography (MDCT), hybrid imaging (positron emission tomography with computed tomography - PET/CT; positron emission tomography with magnetic resonance-PET/MR). In this case report we evaluate usefulness of hybrid imaging (FDG PET/CT) in detection of cardiac sarcoidosis.

Key words sarcoidosis, hybrid imaging

Introduction

Sarcoidosis is a systemic granulomatous disease of unknown cause. Approximately 25% of all patients with sarcoidosis have clinical manifestations of cardiac involvement, whereas myocardial granulomas can be identified in 25–79% of autopsy examinations^{1,2}. Granulomas may involve the pericardium, myocardium and endocardium of both the ventricles as well as the atria².

In diagnosis and clinical assessment of disease activity broad spectrum of analysis is used: clinical examination, laboratory analysis (ACE, chitotriosidase), multi-detector computer tomography (MDCT), hybrid imaging (positron emission tomography with computed tomography- PET/CT; positron emission tomography with magnetic resonance-PET/MR).

Case report

A 59-year-old male complained of prolonged cough and shortness of breath. The patient had no previous history of smoking, alcohol abuse or past surgical experiences. After examination by a pulmonologist, MSCT of the lungs, determination of ACE and biopsy of nodular skin changes, the patient was diagnosed with sarcoidosis and treated with corticosteroids.

Five years after the diagnosis, and after intermittent corticosteroid therapy, the patient presented with exertional dyspnea, with mild limitation during ordinary physical activities. An electrocardiogram revealed a complete left bundle branch block and sinus tachycardia. Doppler echocardiography showed a severely

dilated left ventricle (LV; 64 and 72 mm in systole and diastole, respectively), and a mildly dilated left atrium (41 mm). The LV ejection fraction (LVEF) was 30%. Carvedilol, losartan and furosemide were initiated, with clinical improvement.

In order to assess probable cardiac involvement in sarcoidosis, patient was sent to FDG PET/CT examination. FDG PET/CT examination was performed on a 64-slice hybrid PET/CT scanner (Biograph, Siemens Medical Solutions USA Inc.). Patient fasted for 8 hours before receiving an intravenous injection of 5.50 MBq of 18F-FDG per kilogram. PET/CT acquisitions started 60 min after tracer injection. A 3-dimensional PET scan (6–7 fields of view-, 3 min/field, 30% overlap of beds, the bed width of ~16 cm) and low-dose non enhanced CT scan were acquired from the skull to the mid-thigh.

Low-dose CT, attenuation corrected PET, and combined PET/CT images were displayed for analysis on a Syngo Multi modality workplace (Siemens AG). "Positive" results were above the background level (blood pool and surrounding normal lung parenchyma and/or lymph node activity above the blood pool activity)^{3,4}. Quantitative analysis of FDG uptake in the lesion was done based on the maximum standardized uptake value per focus (SUVmax). This value was calculated as the activity concentration measured at the end of the scan and corrected for individual body weight and dose injected as follows: tissue activity (counts/pixel/s) multiplied by the calibration factor and divided by an injected FDG dose (MBq/kilogram of bodyweight) [3,4]. FDG PET/CT examination revealed cardiac sarcoidosis in this patient, with focal pattern of uptake of FDG in myocardium, SUV max 5.8 (Figure 1).

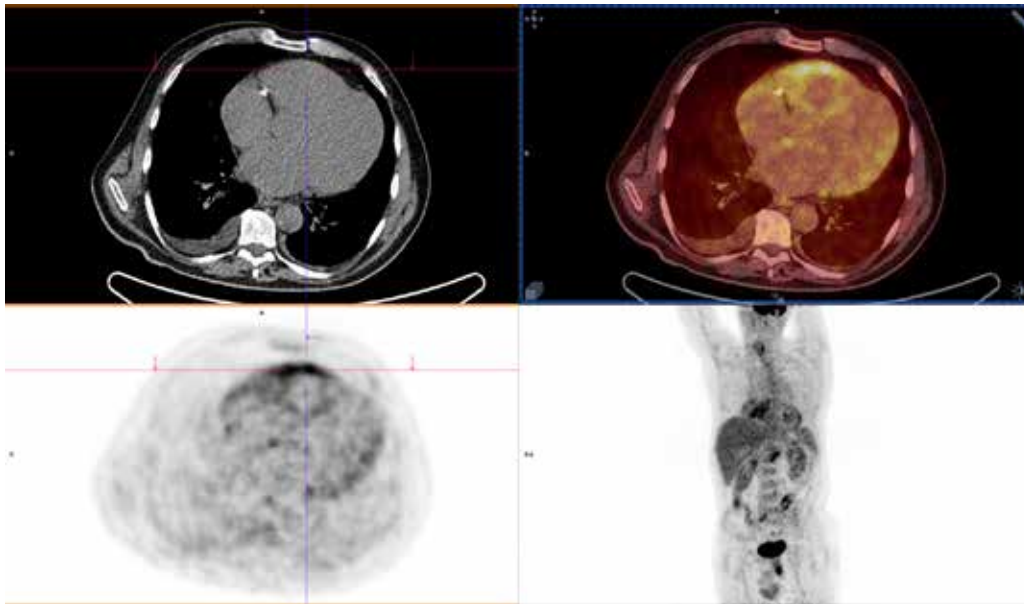


Figure 1. Cardiac sarcoidosis presented as focal FDG myocardial uptake on the FDG PET/CT examination

Discussion

Cardiac involvement in patients with sarcoidosis is being increasingly recognized and is associated with poor prognosis. Although environmental and genetic factors have been implicated in its pathogenesis, the etiology of cardiac sarcoidosis remains obscure.

The myocardium of the left ventricular free wall, particularly at the base of the heart, is most commonly affected, followed by the basal inter ventricular septum. Also, there appears to be a predilection for involvement of the conduction system^{1,2,5}. Granulomas in the ventricular myocardium may lead to abnormal automaticity and re-entrant tachyarrhythmias manifesting as palpitations or syncope. Involvement of the conduction system may lead to bradyarrhythmias and syncope⁵.

Congestive heart failure may result from widespread sarcoidosis of the myocardium with a decline in left ventricular ejection fraction and death².

Diagnosis of cardiac sarcoidosis remains challenging based on a broad and nonspecific set of signs and symptoms ranging from asymptomatic electrocardiographic findings to sudden death and progressively worsening heart failure and arrhythmias^{1,5}.

Cardiac sarcoidosis is definitely confirmed with endomyocardial biopsy^{2,5}. Given the invasiveness and risks of the procedure, other alternative diagnostic methods are used.

Diagnosis and clinical assessment of disease activity usually implies clinical examination with laboratory analysis (ACE, chitotriosidase) and imaging procedures.

MDCT is the most available and consequently most used. However, molecular imaging such as FDG PET/CT or PET/MR are more informative.

The advantage of FDG PET/CT is providing information of precise anatomic localization of the disease and the functional changes at the cellular level. Additionally, the whole-body study protocol is especially important in order to evaluate the extent of this multisystem disease¹.

Hybrid imaging is more expensive than CT scan, but the higher expense is well balanced by its ability to demonstrate activity/inactivity in lesion, which is not possible with morphologic examinations. Radiation burden of a high resolution or a contrast-enhanced CT scan is accompanied by a greater radiation burden than the one necessary in the PET/CT scan⁷. When using a low dose, the mean effective dose was reduced from 8.10 to 5.50 mSv. The blinded analysis of the image quality showed no clinically significant degradation of the lower-dose studies⁷. Further studies using PET/MR may further decrease the radiation dose, and bring additional information about this pathology.

Taken together, for a definitive diagnosis and management all patients with cardiac sarcoidosis will likely benefit from an echocardiogram and either cardiac MR or FDG PET/CT or PET/MR^{2,5}. FDG PET/CT imaging could be performed to establish baseline disease activity, assess need for initiation of medical therapy and monitor response to treatment over time. Alternatively, in patients with contraindication to performing MR, FDG PET/CT could be used as first line imaging.

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Sažetak

Otkrivanje sarkoidoze srca putem hibridnog imidžinga

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Sarkoidoza je sistemska granulomatozna bolest nepoznatog uzroka. Otprilike jedna četvrtina svih pacijenata sa sarkoidozom ima kliničke manifestacije sarkoidoze srca. Granulomi mogu zahvatiti perikard, miokard i endokard obe komore, kao i pretkomore. Sarkoidoza srca je povezana sa lošom prognozom i definitivno se potvrđuje endomiokardijalnom biopsijom. S obzirom na invazivnost i rizike procedure, u dijagnostici srčane sarkoidoze koriste se druge alternativne dijagnostičke metode. U dijagnozi i kliničkoj proceni aktivnosti bolesti koristi se širok spektar analiza: klinički pregled, laboratorijske analize (ACE, hitotriosidaza), multidetektorska kompjuterizovana tomografija (MDCT), hibridne dijagnostičke procedure (pozitronska emisiona tomografija sa kompjuterskom tomografijom - PET / CT; pozitronska emisiona tomografija sa magnetnom rezonancom-PET / MR). U ovom prikazu slučaja biće prikazana korisnost FDG PET / CT u otkrivanju srčane sarkoidoze.

Ključne reči: sarkoidoza, hibridni imaging