Cardiac Sarcoidosis versus Arrythtmogenic Right Ventricular Dysplasia/ Cardiomyopathy

Andrés Ricardo Pérez-Riera MDPhD.

Patients with cardiac sarcoidosis may present with clinical and morphological features similar to ARVC/D or cardiomyopathy (Ott 2003). Sarcoidosis is an inflammatory granulomatosis entity of unknows cause, characterized by multisystemic involvement. Practically no organ is immune to sarcoidosis; most commonly, in up to 90% of patients, it affects the lungs. (Hoitsma 2004). The most commonly involved organ in sarcoid related death has been reported to be the lung in western countries, while it was the heart in the Japanese autopsy series. (Iwai 1994).

The diagnosis of myocardial sarcoidosis is difficult and frustrating.Its clinical manifestations depend on the location and extentof granulomatous inflammation, and the symptoms and signs rangeamong benign arrhythmias, heart block, intractable CHF,intense chest pain, to fatal VF. (Sharma 2003).

The ECG finding may be normal or may reflect every degree ofblock of the atrioventricular junction and bundle of His and every type of arrhythmia along with nonspecific ST-T-wavechanges (Flemming, 1994).

Cardiac sarcoidosis should be considered in all young patients with unexplained conduction disorders,(Kollermann 2001) CHF or in cases of SCD (Lip 1996).

In extensive forms are frequently pseudo myocardial infarction patterns with pathological Q waves on ECG. (Shindo 1998).

MRI abnormalities, consisting of cardiac signalintensity and thickness, with the following threepatterns:

1) Nodular;
2) Focal increase in signal on gadolinium diethylenetriaminepentaacetic acid-enhanced, T1-weighted images;
3) Focal increasedsignal on T2-weighted images without gadolinium uptake.

The improvement or stabilityof the MRI findings is correlated with clinical features.

With corticosterotherapy, the MRIimages improved either partially or completely, whereas.

The cardiac MRI may find its usefulness as a guide toobtaining EMB specimens and to monitoringthe response of the disease to treatment.

The study issmall and lacks a correlation of myocardial histology with MRIfeatures. However, the study clearly calls for a large multicentertrial.

The most significantdrawback of MRI is that the patient with a pacemaker and/ orautomatic ICD will not beable to take advantage of it. In such patients, ^{201}Tl scanningremains the test for assessing myocardial damage.

Cardiac PET using (18) F-FDG under fasting conditions (fasting (18) F-FDG PET) is a promising technique for identification of cardiac sarcoidosis and assessment of disease activity. The methodology can detect the early stage of cardiac sarcoidosis, in which fewer perfusion abnormalities and high inflammatory activity are noted, before advanced myocardial impairment. The sensitivity of fasting (18) F-FDG PET in detecting cardiac sarcoidosis was

100%, significantly higher than that of (99m)Tc-MIBI SPECT (63.6%) or (67)Ga scintigraphy (36.3%). The accuracy of fasting (18) F-FDG PET was significantly higher than (67) Ga scintigraphy.(Okumura 2004.).
An EMB is preferable, but the procedure has sensitivity aslow as 20% (*Uemura, 1999*). Others author referred sensitivity approximately of 50% thus, the search for a safe, reliable, and easilyavailable diagnostic test for cardiac sarcoidosis continues. The pathological feature is the presence of noncaseating granulomas that eventually form fibrotic scars.
The table below shows the principal differences between the two entities:

	Cardiac Sarcoidosis	ARVC/D
Family history:	Absent.	Present in 30% to 50% of cases. When the disease is identified genetic screening should be performed among patient's family members.
Gender (M/F):	1 to 1.	2.9 to 1
Age at presentation:	Young or middle-aged adults.	Adolescents and young adults, perhaps There are rare references in childhood
Multisystemic involvement:	Yes.	No.
Chest pain:	Intense chest pain is refered.	No.
Clinical myocardial restrictives features:	Posible.	No.
Mitral regurgitation:	Is common.	Only in late estage with involvement of LV.
Pseudo myocardial infarction patterns on ECG:	Fequent in extensive forms.	No.
Chest roentgenogram:	Bilateral hiliar linphoadenopathy.	Eventually RV cardiomegaly.
Lungs affectation:	In up to 90% of patients. Cor pulmonale is frequent.	No.

Pathological features:	Noncaseating granulomas that eventually form fibrotic scars.	Typical fibro-fatty replacement of the RV myocardium on dysplasia triangle.
More comum cardiac sites involved:	LV free wall and interventricular septum.	RVOT, RVIT, and apex of RV.
Pericardial effusion:	Are not uncommon.	Absent.
Improved MRIimages with corticosteroids:	Yes.	No.
Therapy with corticosteroids, hydroxychloroquine, methrotexate or cyclophosphamide:	Sometime are indicated. (Mitchell 1997). Immunosuppressive and anticytokine treatments can be effective in severe systemic sarcoidosis and should be considered in sight-threatening disease.	No.

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