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Differential diagnosis between ARVD and cardiac sarcoidosis

Andrés Ricardo Pérez Riera[1] and Augusto Hiroshi Uchida[2]

[1]Electro-Vectocardiology Sector of the Discipline of Cardiology, ABC Faculty of Medicine (FMABC), Foundation of ABC (FUABC), Santo André, Sao Paulo, Brazil
[2]Medical Attendant, Exercise Testing Division Heart Institute, University of Sao Paulo Medical School, Sao Paulo, Brasil

Patients with cardiac sarcoidosis may present with clinical and morphological features similar to ARVD or cardiomyopathy [1]. Sarcoidosis is an a chronic noncaseating granulomatous disease of unknown cause, characterized by multisystemic involvement. Practically no organ is immune to sarcoidosis; most commonly, in up to 90% of patients, it affects the lungs [2]. 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 [3].

The diagnosis of myocardial sarcoidosis is difficult and frustrating. Its clinical manifestations depend on the location and extent of granulomatous inflammation, and the symptoms and signs range among benign arrhythmias, heart block, intractable CHF, intense chest pain, to fatal VF [4].

The ECG may be normal or reflect every degree of block of the atrioventricular junction and bundle of His and every type of arrhythmia along with nonspecific ST-T-wave changes [5]. Other common ECG findings are signs of hypercalcemia (e.g. decreased QT interval), PR depression caused by pericarditis, or ST elevation caused by ventricular aneurysm. Premature ventricular complexes and nonsustained ventricular tachycardia are also common, seen by ECG in as many as 22% of patients with sarcoidosis. Sudden death from ventricular tachycardia and heart block accounts for up to 65% of deaths from cardiac sarcoidosis [6].

Address for correspondence: Andrés Ricardo Pérez Riera, MD
Electro-Vectocardiology Sector of the Discipline of Cardiology
ABC Faculty of Medicine (FMABC)
Foundation of ABC (FUABC)
Santo André, Sao Paulo, Brazil
e-mail: riera@uol.com.br, augusto.uchida@incor.usp.br

Cardiac sarcoidosis should be considered in all young patients with unexplained conduction disorders [7], CHF or in cases of SCD [8].

In extensive forms are frequently pseudo myocardial infarction patterns with pathological Q waves on ECG [9].

MRI abnormalities, consisting of cardiac signal intensity and thickness, with the following three patterns:

— nodular;
— focal increase in signal on gadolinium diethylenetriamine pentaacetic acid-enhanced, T1-weighted images;
— focal increased signal on T2-weighted images without gadolinium uptake.

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

With corticosterotherapy, the MRI images improved either partially or completely, whereas.

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

The study is small and lacks a correlation of myocardial histology with MRI features. However, the study clearly calls for a large multicenter trial.

The most significant drawback of MRI is that the patient with a pacemaker and/or automatic ICD will not be able to take advantage of it. In such patients, ^{201}Tl scanning remains the test for assessing myocardial damage.

Cardiac PET using F-FDG under fasting conditions is a promising technique for identification of cardiac sarcoidosis and assessment of disease activity. The metodology 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 FDG PET in detecting cardiac

sarcoidosis was 100%, significantly higher than that of ^{99m}Tc-MIBI SPECT (63.6%) or Ga scintigraphy (36.3%). The accuracy of fasting FDG PET was significantly higher than Ga scintigraphy [10].

An EMB is preferable, but the procedure has sensitivity as low as 20% [11]. Others author referred sensitivity approximately of 50% thus, the search for a safe, reliable, and easily available diagnostic test for cardiac sarcoidosis continues. The pathological feature is the presence of noncaseating granulomas that eventually form fibrotic scars. Table 1 shows the principal differences between the two entities.

Table 1. The principal differences between the two entities.

	Cardiac sarcoidosis	**ARVD**
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. Sarcoidosis commonly involves the eye, causing uveitis, the lacrimal gland, and the cranial nerves, including the optic nerve itself [12].	No.
Chest pain	Intense chest pain is referred.	Sometime [13–15]. Based on the data of Chinese ARVD registry, it is not uncommon that ARVD patients have chest pain.
Clinical myocardial restrictives features	Possible.	No.
Mitral regurgitation	Is common.	Only in late stage with involvement of LV.
Pseudo myocardial infarction patterns on ECG	Frequent in extensive forms.	No.
Chest roentgenogram Pathological features	Bilateral hiliar linphoadenopathy. Noncaseating granulomas that eventually form fibrotic scars. From the histological studies, fat infiltration seems absent in cardiac sarcoidosis.	Eventually RV cardiomegaly. Typical fibro-fatty replacement of the RV myocardium on dysplasia triangle.
Lungs affectation	In up to 90% of patients. Cor pulmonale is frequent.	No.
More comum cardiac sites involved	LV free wall and interventricular septum.	RVOT, RVIT, and apex of RV.
Pericardial effusion	Are not uncommon.	Absent.
Improved MRI images with corticosteroids	Yes.	No.
Extensive angiographic analysis. With right ventricular angiography	Ventricular aneurysm Are sometime present [13]. Free wall of the right ventricle thin, dyskinetic with dilation is possible [16].	The best definition of ARVD is obtained by extensive angiographic analysis with measurement of oxygen saturation and pressure curves in different positions, coronary angiography and biventricular angiography in order to distinguish between some in regard to right ventricular involvement similar cardiac entities [17].
Therapy with corticosteroids, hydroxychloroquine, methrotexate or cyclophosphamide	Sometimes are indicated [18]. Immunosuppressive and anticytokine treatments can be effective in severe systemic sarcoidosis and should be considered in sight-threatening disease.	No.

References

1. Ott P, Marcus FI, Sobonya RE et al. Cardiac sarcoidosis masquerading as right ventricular dysplasia. Pacing Clin Electrophysiol, 2003; 26: 1498–1503.
2. Hoitsma E, Faber CG, Drent M, Sharma OP. Neurosarcoidosis: a clinical dilemma. Lancet Neurol, 2004; 3: 397–407.
3. Iwai K, Sekiguti M, Hosoda Y. Racial difference in cardiac sarcoidosis incidence observed at autopsy. Sarcoidosis, 1994; 11: 26–31.
4. Sharma OP. Diagnosis of cardiac sarcoidosis an imperfect science. A Hesitant Art Chest, 2003; 123: 18–19.
5. Flemming H, Bailey S. Cardiac sarcoidosis. In: James DG eds. Sarcoidosis and other granulomatous disorders. Vol. 73. Marcel Dekker, New York 1994; 323–334.
6. Roberts WC, McAllister HA Jr, Ferrans VJ. Sarcoidosis of the heart. A clinicopathologic study of 35 necropsy patients (group 1) and review of 78 previously described necropsy patients (group 11). Am J Med, 1977; 63: 86–108.
7. Kollermann J, Roos G, Helpap B. Sudden cardiac death from unrecognized cardiac sarcoidosis Pathologe, 2001; 22: 141–144.
8. Lip GY, Gupta J, Gill JS et al. Sarcoid heart disease: a rare cause of chest pain and malignant cardiac arrhythmia in a young Asian man. A case report. Angiology, 1996; 47: 905–910.
9. Shindo T, Kurihara H, Ohishi N et al. Images in cardiovascular medicine. Cardiac sarcoidosis. Circulation, 1998; 97: 1306–1307.
10. Okumura W, Iwasaki T, Toyama T, Usefulness of Fasting 18F-FDG PET in Identification of Cardiac Sarcoidosis. J Nucl Med, 2004; 45: 1989–1998.
11. Uemura, A, Morimoto, S, Hiramitsu S et al. Histologic diagnostic rate of cardiac sarcoidosis: evaluation of endocardial biopsies. Am Heart J, 1999; 138: 299–302.
12. Jones NP. Sarcoidosis. Curr Opin Ophthalmol, 2002; 13: 393–396.
13. Anderson EL. Arrhythmogenic right ventricular dysplasia. Am Fam Physician, 2006; 73: 1391–1398.
15. Peters S, Trummel M, Meyners W. Prevalence of right ventricular dysplasia-cardiomyopathy in a nonreferral hospital. Int J Cardiol, 2004; 97: 499–501.
16. Hulot JS, Jouven X, Empana JP, Frank R, Fontaine G. Natural history and risk stratification of arrhythmogenic right ventricular dysplasia/cardiomyopathy. Circulation, 2004; 110: 1879–1884.
17. Marks A, Anderson MH, Harrison NK. Ventricular aneurysm secondary to sarcoid disease. Heart, 2004; 90: 694.
18. Shiraishi J, Tatsumi T, Shimoo K et al. Cardiac sarcoidosis mimicking right ventricular dysplasia. Circ J, 2003; 67: 169–171.
19. Peters S, Weber B, Hartwig CA et al. Incidence and significance of morphologic and morphometric variants in selective right ventricular angiography in diagnosis of arrhythmogenic right ventricular disease Med Klin (Munich), 1994; 89: 175–183.
20. Mitchell DN, du Bois RM, Oldershaw PJ. Cardiac sarcoidosis. BMJ, 1997; 314: 320–321.