May Active Myocarditis Mimick Arrhythmogenic Right Ventricular Cardiomyopathy Phenotype by Electroanatomic Mapping?

We read with interest the study of Pieroni et al. (1). Although we fully agree on the superiority of endomyocardial biopsy (EMB) for a specific diagnosis of the underlying disease, especially considering that fibro-fatty replacement and low electrical voltage also may occur in the setting of sarcoid cardiomyopathy (2), we have some concerns with regard to their findings.

The authors report that 50% of patients with the clinical diagnosis of arrhythmogenic right ventricular cardiomyopathy (ARVC) had “active myocarditis.” This means that an ARVC phenotype (inverted T-wave in right pre-cordial leads, ventricular tachycardia of left bundle branch block morphology, morphological abnormalities of the right ventricle at echocardiography/angiography/magnetic resonance imaging [MRI], late potentials, >2,000 premature ventricular beats at Holter monitoring) frequently corresponds to myocarditis with predominant involvement of the right ventricle. In our longstanding experience with EMB, this is a rare occurrence. Additionally, 30 patients with an ARVC phenotype collected in 1 year seem unrealistic for a single center.

An inflammatory infiltrate of 7 lymphocytes/mm² is too scanty to be considered diagnostic for active myocarditis. This number is within the normal limits. In all the published literature on voltage mapping, none have previously reported a positive voltage map with “active myocarditis” (3,4). Only a phenomenon of scarring associated with myocarditis may account for such a loss of myocardium to generate low electrical voltage, and this may occur in the setting of chronic (nonacute) myocarditis.

Although it has not been documented, MRI did not show significant differences between patients with “active myocarditis” and ARVC in terms of delayed enhancement, which was high both in the right and in the left ventricular walls, a finding again suggestive of extensive replacement-type fibrosis in both subgroups. Surprisingly, the authors did not find any evidence of fibrosis at EMB in any patient with “active myocarditis,” positive electroanatomic mapping, and delayed enhancement at MRI.

It is noteworthy that the patient in Figure 2 of Pieroni et al. (1), with positive electroanatomic mapping and a histological diagnosis of active myocarditis, shows a clear-cut picture at angiography of “pile d’assiettes,” which is almost pathognomonic of ARVC (5).

*Gaetano Thiene, MD
Cristina Basso, MD, PhD
Domenico Corrado, MD, PhD

“pile d’assiettes,” which is almost pathognomonic of ARVC (5).

of active myocarditis, shows a clear-cut picture at angiography of

with positive electroanatomic mapping and a histological diagnosis

extensive replacement-type fibrosis in both subgroups. Surprisingly,

right and in the left ventricular walls, a finding again suggestive of

ARVC in terms of delayed enhancement, which was high both in the

right and in the left ventricular walls, a finding again suggestive of

extensive replacement-type fibrosis in both subgroups. Surprisingly,

the authors did not find any evidence of fibrosis at EMB in any

patient with “active myocarditis,” positive electroanatomic mapping,

and delayed enhancement at MRI.

It is noteworthy that the patient in Figure 2 of Pieroni et al. (1),

with positive electroanatomic mapping and a histological diagnosis

of active myocarditis, shows a clear-cut picture at angiography of

“pile d’assiettes,” which is almost pathognomonic of ARVC (5).

*University of Padua
Pathological Anatomy
Via A. Gabelli, 61
35121 Padova
Italy
E-mail: gaetano.thiene@unipd.it

REFERENCES


Reply

We appreciate the interest of Dr. Thiene and colleagues in our work (1). However, despite their longstanding experience in this field, some of their comments could cause a misleading interpretation of our findings.

In particular, their concern regarding the prevalence of right ventricular myocarditis among patients with a diagnosis of noninvasive arrhythmogenic right ventricular cardiomyopathy (ARVC) is surprising when considering that in a recent study (2) they diagnosed an active myocarditis in 40% of patients with a clinical diagnosis of ARVC (10 of 25 patients with a definite histological diagnosis). As well as Padua University, our institution is a referral center for arrhythmias and cardiomyopathies, and a significant proportion of the patients enrolled in 1 year were referred for further invasive studies from southern Italian hospitals after noninvasive evaluation suggested a diagnosis of ARVC.

With regard to histological evaluation, we clearly stated that the diagnosis of active myocarditis was based on Dallas criteria but also required the demonstration of a significant number of activated and cytotoxic T lymphocytes. An amount of >7 lymphocytes/mm², even if identified as activated/cytotoxic, can be still considered arbitrary, as it is the case for other criteria adopted by other groups (3). Moreover, confusion should be avoided between the terms “active” and “acute” representing, respectively, a histological and clinical definition. It is evident from the text and tables that all