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Editorial

Another small puzzle card in the cardiac involvement due to autoimmune diseases

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In the present issue of the International Journal of Cardiology Mavrogeni et al. [1] addressed the relevant topic of arrhythmias in patients affected by various rheumatic disorders. As clinicians know, the abrupt onset of arrhythmias might severely complicate the clinical picture of these patients despite a persistent normal regional and global left ventricular function. In fact, from the data shown, it appears that macroscopic changes such as ventricular dysfunction and, in this population, even macroscopic accumulation of contrast agent (Late Gadolinium Enhancement - LGE) are not related to the appearance of arrhythmias, while an abnormal enlargement of the interstitial space (Extra Cellular Volume - ECV) seems to be related to electrical instability. These findings are not surprising if we consider the intrinsic nature of rheumatic disorders, where a likely persistent inflammatory process takes place, possibly quite early in the history of the disease. Recently, other studies have linked the presence of LGE to rhythm disturbances [2] or even to cardiovascular events [3], suggesting a possible role of CMR to identify those patients more prone to adverse events. This is relevant also because rheumatic patients may have more contraindications to some antiarrhythmic therapies.

The second message which can be derived from this paper is that not all the rheumatic diseases have the same risk profile, being systemic sclerosis the potentially more dangerous. There are preliminary reports on the usefulness of implanting an ICD in systemic sclerosis (SSc) patients with ventricular arrhythmias, even when not reaching full indications according to current guidelines [4]. Patients with autoimmune disease have chronic, systemic, often progressive conditions that may need different criteria compared to patients with other heart disease. If these findings will be confirmed by robust data, the clinician might be able to discriminate between patients at low and high risk to develop ventricular arrhythmias, with the possibility of modulating the therapy and, in selected cases, even to discuss the cost/benefits of implanting an ICD for primary prevention.

On the other side, the possibility to evaluate microstructural abnormalities such as ECV in a non-invasive and repeatable approach, such as by CMR, for such a relevant problem represents a new concept on the way to understand the course of these diseases, especially at early stages.

It is not difficult to foresee a systematic use of CMR (LGE, T1 mapping, ECV) in these patients, similarly to what is happening in other conditions, where the involvement of the heart is known but not yet fully understood such as accumulation disorders (ex. thalassemia, Anderson-Fabry, Amyloidosis, etc.).

If these parameters will be used, not as a surrogate of endomyocardial biopsy [5], but as a relevant point in the decision-making together with the other clinical, biohumoral and instrumental findings, the clinician will have the new possibility to monitor the degree of the often subclinical silent burning of cardiac involvement, and hopefully to prevent adverse events, which so far is still unpredictable.

Finally, the methodology proposed is worth of further discussion. T1 mapping and derived parameters are still under development and there is the substantial risk of lack of reproducibility between different labs, different acquisition procedures, different scanners, analysis of data, availability of normal datasets, etc. Despite these limitations, which are prevalently linked to the novelty of the methodology itself, the data shown in this paper are quite encouraging and can serve as a stimulus to invest toward this direction.

Conflict of interest

No conflict of interest has to be disclosed.

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