EDITORIAL

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Can documented coronary vasospasm be the smoking gun in settling the etiology of sudden cardiac death?

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Sudden cardiac death is one of the leading causes of mortality. Despite improvements in the emergency transportation system and the introduction of a novel resuscitation process, only a small proportion of involved patients have favorable clinical outcomes [1]. As a result, the community is still burdened with great socioeconomic sequelae despite grappling with this staggering complex problem. Furthermore, a substantial proportion of patients experience recurrent events, even with optimal medical care. Therefore, identifying the etiology of sudden cardiac death is of great importance in the management of this event.

Inherited primary arrhythmia syndrome (IPAS) is an electrical abnormality caused by functional derangement of the cardiac ion channel. It presents as various phenotypes according to the involved genetic abnormality, i.e., congenital long QT syndrome, Brugada syndrome, early repolarization syndrome, and catecholaminergic polymorphic ventricular tachycardia. IPAS accounts for a significant proportion of sudden cardiac deaths, especially in young, healthy individuals. The diagnosis of IPAS is principally made by documentation of characteristic electrocardiogram (ECG) findings; however, the ECG presentation usually has significant temporal variations, and typical ECG findings can appear transiently in many cases, which is a major obstacle in the diagnosis of IPAS [2,3].

Vasospastic angina has been recognized as a major cause of ischemic heart disease since it was first described by Prinzmetal et al. [4]. Although it is generally considered a low-risk disease, devastating clinical events, such as ventricular arrhythmia and subsequent sudden cardiac death, can occur. In one large registry of cardiac arrest survivors, a considerable number of patients with apparently unexplained cardiac arrest exhibited evidence of coronary spasm [5]. Therefore, when determining the etiology of aborted sudden cardiac death, the evaluation of coronary vasospasm should be considered and may be mandatory if there is no other apparent cause. However, angiographically-proven coronary vasospasm is not sufficient to exclude other possible etiologies. A vasodilator therapy that includes a calcium channel blocker and nitrate is usually quite effective for the prevention of recurrent vasospasm, but a substantial proportion of patients experience recurrent devastating events despite adequate

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medical therapy [6]. These events are associated with drug refractoriness and other factors, such as failure to cease smoking, but other underlying masked etiologies should also be considered [7].

In this issue, Lee at al. [8] report that IPAS can be masked in patients diagnosed with vasospastic angina. In this study, 60.8% of patients with proven coronary vasospasm had typical findings of IPAS: Brugada-type patterns in six (8.2%), arrhythmogenic right ventricular dysplasia patterns in three (4.1%), long QT syndrome pattern in one (2.2%), and early repolarization in 38 (51.4%). They also reported a strikingly high rate of recurrent sudden cardiac death (35.6% of enrolled patients) in patients with evidence of coexisting IPAS during a median follow-up duration of 3.9 years.

The coexistence of vasospasm and IPAS has been reported previously [9]; this may be a coincidence, but there is a pathophysiological link between the two disease entities. Experimental data support that temporal and regional heterogeneity of ventricular repolarization are an important pathomechanism of IPAS [10]. Additionally, myocardial ischemia can trigger the development of ventricular arrhythmia in affected persons by increasing heterogeneity. According to a previous report, the coexistence of anomalous coronary artery origin can represent a triggering factor in patients with congenital long QT syndrome [11]. Another report demonstrated that only mild ischemia and vagal influence induced by coronary vasospasm could precipitate ventricular fibrillation in patients with Brugada syndrome [12]. Overlooking underlying IPAS can result in a devastating event, i.e., recurrent ventricular arrhythmia and sudden cardiac death. The strikingly high incidence of recurrent cardiac death in this study supports this remark.

There are clinical implications of great importance and lessons to learn from this study. IPAS can be frequently masked and overlooked in patients with sudden cardiac death. It is necessary to identify the underlying channelopathy in patients with sudden cardiac death, even when coronary spasm is proven spontaneously or by a provocation test.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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