Journal of Electrocardiology 55 (2019) 16–19

Contents lists available at ScienceDirect

Journal of Electrocardiology

journal homepage: www.jecgonline.com

Is the spiked helmet sign the manifestation of long QT syndrome?

András Simon, MD *, Zoltán Járai, MD, PhD

Department of Cardiology, St. Imre University Teaching Hospital, Tétényi út 12-16, 1115 Budapest, Hungary

ARTICLE INFO

Keywords:
Spiked helmet sign
Macroscopic T-wave alternans
Long QT syndrome
Takotsubo syndrome

ABSTRACT

We present the case of a patient with Takotsubo syndrome developing simultaneous inferior, anterior spiked helmet sign (SHS) and macroscopic T-wave alternans (TWA) leading to torsade de pointes ventricular tachycardia (TdP VT). Based on our observations we propose that the SHS is a type of manifestation of critically prolonged QT(U).

Case report

We present the case of a 47-years-old homeless woman with a history of smoking, alcohol abuse and toe amputations due to Buerger's disease. She was admitted to the emergency department with several days' history of recurrent chest pain, shortness of breath, repeated collapses and convulsion. She was wheezing and had acrocyanosis. CT scan of the brain showed chronic vascular encephalopathy with an old lacunar infarct, while chest CT scan ruled out pulmonary embolism. High sensitivity troponin I was elevated (224 ng/L, reference: <13 ng/L), therefore she was referred to the department of cardiology with myocardial infarction. At this point her previously obtained ECGs were reevaluated by a cardiologist. These showed SHS both in the inferior and anterior leads, in addition to long QT and macroscopic TWA (Figs. 1, 2). During the cardiologist's examination at the emergency department serial, short runs of TdP VT was seen on the monitor followed by the patient's short convulsions (Fig. 3). These episodes might have been previously falsely evaluated as an artifact due to the patient's movement during her prior monitoring. After intravenous administration of potassium and magnesium the TdP VT runs and subsequent convulsions stopped within minutes. Bedside echocardiography revealed circumferential akinesis of the mid and apical segments of the left ventricle with moderately decreased global systolic left ventricular function. The wall motion abnormality extended beyond the territory of one single coronary artery, which is characteristic of Takotsubo syndrome (Fig. 4). Repeated troponin level did not rise, therefore coronary angiography was performed only a few days later. This showed normal coronary arteries. After further treatment, the patient made a good recovery. Repeated echocardiography 12 days later showed good left ventricular systolic function and no wall motion abnormality. At this time the ECG demonstrated the regression of the initial abnormalities. Retrospective, aimed questioning revealed that the patient underwent significant emotional stress prior to her admission following the separation from her partner. All together we found the diagnosis of Takotsubo syndrome well established.

Discussion

The first description of SHS drew attention to a new ECG marker, which can deceivingly mimic ST-segment elevation myocardial infarction (STEMI).

In the cases originally presented, the SHS was present only in the inferior leads characteristically with an ST-segment elevation associated with an upward baseline shift starting before the onset of the QRS complex. Each patient had a critical non-cardiac illness associated with high in-hospital mortality, but never a STEMI [1].

Since the first description, numerous other reports have been published where the SHS was present in the chest leads, but not in the inferior leads. The exact cause and pathomechanism of the SHS has not been clarified so far. Some observations recognized that SHS in the inferior leads was accompanied with acute abdominal events and when it was seen in the chest leads, an acute thoracic event was indicated. For this reason, it was hypothesized that it might be in connection with an acute rise in the intraabdominal or intrathoracic pressure [2]. However, other cases do not support these hypotheses as SHS was seen in cerebral catastrophes [3] and also after stellate ganglion ablation [4]. Although the latter cases were not in connection with abdominal or thoracic events, a common mechanism may be significant adrenergic excess.

* Corresponding author.
E-mail address: dr.andras.simon@gmail.com (A. Simon).

https://doi.org/10.1016/j.jelectrocard.2019.04.011
0022-0736/

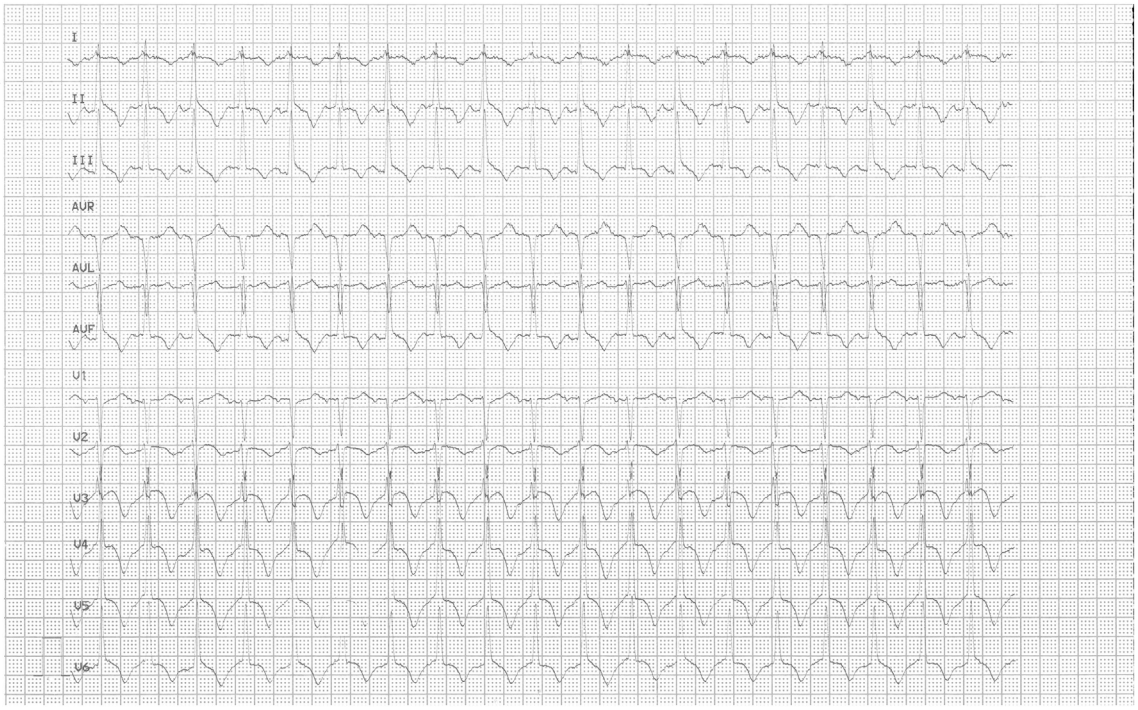

Fig. 1. Initial electrocardiogram obtained at the emergency department showing inferior and anterior spiked helmet sign and macroscopic T-wave alternans.

To our best knowledge the simultaneous appearance of macroscopic TWA and SHS both in the inferior and anterior leads has not yet been reported.

Macroscopic TWA, refers to the beat-to-beat alternation of the size or polarity of the T-wave. It is rarely seen on 12-lead ECG. Conventionally it is held to be the sign of electric instability and preceding life threatening malignant ventricular arrhythmia, especially imminent TdP VT, and is frequently associated with congenital or acquired QT prolongation. It is known, that Takotsubo cardiomyopathy can cause QT prolongation and macroscopic TWA has been recently reported in Takotsubo syndrome [5].

Just like macroscopic TWA, the SHS is another marker that might draw attention to the underlying, but not necessarily always evident long QT syndrome.

Laundon's subsequent hypothesis explains the SHS with the deformation of the baseline by the grossly prolonged repolarization of the previous cardiac cycles, where the upward shift represents the previous giant T-U-wave, which overlaps the QRS complexes [3].

In this report we propose another way of the evolvement of SHS. Our hypothesis is that every clinical state can create the characteristics of typical SHS, when the QT(U) is long enough and the heart rate is fast enough for the inverted T(U)-wave to reach the following QRS complex. In these cases the downward and upward 'shift' of the baseline are the descending and ascending limbs of the wide, inverted T-waves.

As it might be caused by the coincidence of the above mentioned conditions, it is worth considering this kind of SHS as an 'optical illusion'. Either way - if the 'helmet' in the SHS is formed by the giant positive U-waves extending beyond the next QRS, or is an 'optical illusion' created by the wide, negative T-waves reaching the next depolarization - the question still stands: why are they in connection sometimes with acute critical abdominal and thoracic syndromes?

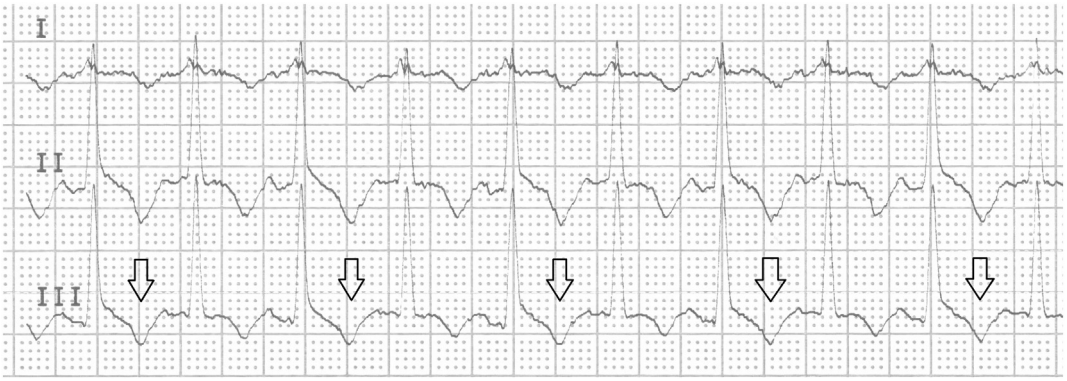

Fig. 2. Enlarged standard leads. Arrows pointing to the alternating T-waves.

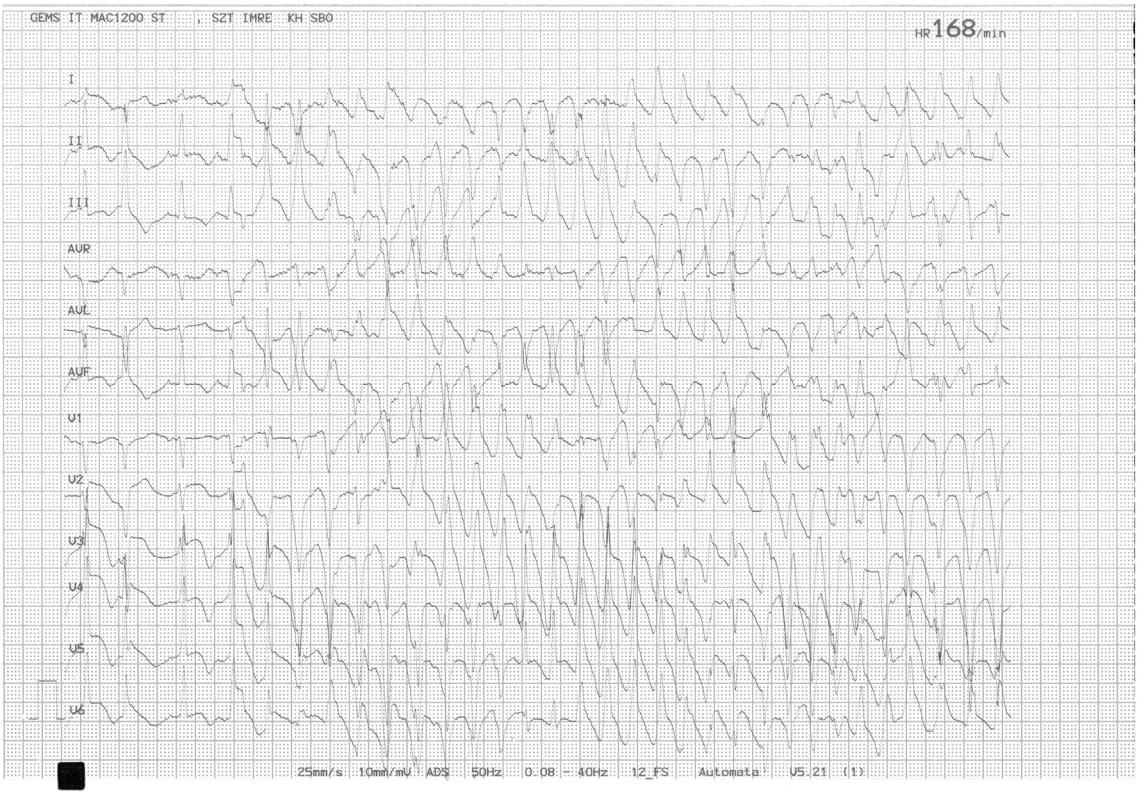

Fig. 3. Torsade de pointes ventricular tachycardia. The ECG was obtained during the examination of the cardiologist at the emergency department.

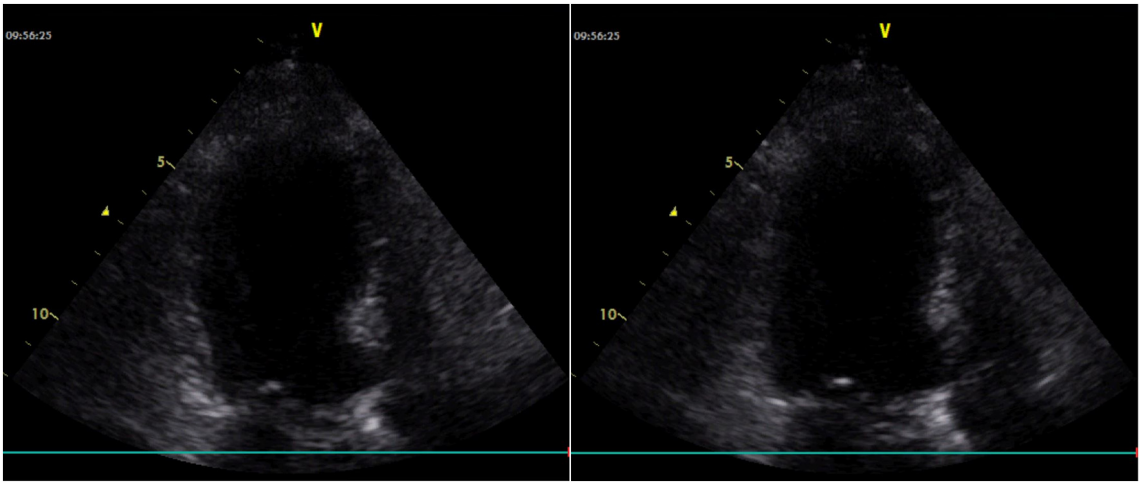

Fig. 4. Echocardiography showing circumferential akinesis of the mid and apical segments of the left ventricle with moderately decreased global systolic left ventricular function. (Apical four chamber view of the left ventricle. Left side: systole, right side:diastole.)

In most cases of SHS, the factors behind the QT prolongation are evident, such as severe electrolyte disturbances, cerebral catastrophes, hypoxia, drug effects or congenital long QT [1–4,6]. In others, Takotsubo syndrome [7] might be a potential connection and explanation, as it can be associated with both critical illness and prolonged QT and wide, inverted T-waves [8].

Summary

The SHS is reputed to be the hallmark of critical illness, but its exact mechanism remains to be clarified. Our present observation supports the theory, that SHS is the overlap of the QRS complex and the prolonged repolarization of the previous cardiac cycle. Not only markedly delayed, giant, positive U-waves can create this phenomenon. If the QT(U) is long enough at a given heart rate, and there is a wide, inverted T-wave, which reaches the next QRS, the scenario will all together look like a typical SHS. Recognition of the spiked helmet pattern may facilitate the identification of the underlying, not always evident QT(U) prolongation and coexisting repolarization abnormality. We believe that Takotsubo syndrome accompanying non-cardiac critical illnesses may explain why SHS is seen in the above-mentioned emergencies.

Declarations of interest

None.

References

[1] Littmann L, Monroe MH. The “spiked helmet” sign: a new electrocardiographic marker of critical illness and high risk of death. Mayo Clin Proc 2011;86(12):1245–6.
[2] Tomcsányi J, Frész T, Proctor P, Littmann L. Emergence and resolution of the electrocardiographic spiked helmet sign in acute noncardiac conditions. Am J Emerg Med 2015;33(1):127 [e125-127].
[3] Laundon RK, Littmann L. Spiked helmet pattern ST elevation in subarachnoid hemorrhage. J Electrocardiol 2019;52:96–8.
[4] Aliyev F, Abdulkerimov V, Gul EE, Samedov F, Isayev E, Ferecov E. Spiked helmet sign after percutaneous left stellate ganglion ablation in a patient with long QT syndrome. J Electrocardiol 2017;50(6):944–6.
[5] Warraich HJ, Buxton AE, Kociol RD. Macroscopic T-wave alternans in a patient with takotsubo cardiomyopathy and QT prolongation. Heart Rhythm Off J Heart Rhythm Soc 2014;11(10):1848–9.
[6] Gomez-Dominguez R, Hidalgo R, Garcia-Rubira JC. Severe hypocalcemia masquerading as acute coronary syndrome. J Emerg Med 2013;45(5):715–7.
[7] Samadov F, Gasimov E, Aliyev F, Isayev E. The “spiked helmet” sign - a potential relationship to Takotsubo cardiomyopathy. Am J Emerg Med 2018;36(2) [345 e345-345 e347].
[8] Pergolini A, Zampi G, Pontillo D, Venturini E, Pino PG, Musumeci F. Atypical recurrence of Takotsubo syndrome: giant T-wave inversion and huge QTc prolongation. J Cardiovasc Med (Hagerstown) 2017;18(8):631–2.