Impact of differences in embryonic origin on the electrical features of the giant right atrium in an elderly patient

Toshihiko Goto, MD,* Kento Mori, MD,* Yoshimasa Murakami, MD,† Nobuyuki Ohte, MD*

From the *Department of Cardiology, Nagoya City University Graduate School of Medical Sciences, Nagoya, Japan, and †Department of Cardiovascular Medicine, Nagoya East Medical Center, Nagoya, Japan.

Introduction

Giant right atrium (RA) is a rare congenital disease. However, the electrical features of giant RA have not been fully elucidated. Here, we present a patient who was suspected of having a giant RA based on his medical history.

Case report

A 79-year-old man was referred to our cardiology department owing to atrial tachycardia and peripheral edema. Physical examination findings indicated right-sided heart failure and tricuspid regurgitation. A 12-lead electrocardiogram revealed inverted flutter waves with a 3:1 conduction in the inferior wall leads and a stable ventricular rate of 75 beats/min. This patient had previously been suspected of having dextrocardia since his 20s based on chest radiography, which showed a deviation of the heart towards the right with a cardiothoracic ratio of 0.55 (Supplemental Figure 1). Echocardiography and cardiac computed tomography (CT) scans (Figure 1A) revealed a dilated RA. No common causes of RA enlargement, such as shunt disease, pulmonary hypertension, tricuspid stenosis, or Ebstein anomaly, were found. He had no family history of heart disease, which included his parents and a daughter in her 40s. The electrophysiology study with the CARTO Electroanatomical Mapping System (Biosense Webster Inc, Irvine, CA) showed a macroreentry with counter-clockwise rotation in the RA tricuspid annulus (Supplemental Video 1). Radiofrequency ablation for the cavotricuspid isthmus was performed using a SmartTouch SF ablation catheter (Biosense Webster), and the atrial flutter was terminated. Voltage mapping during sinus rhythm showed a longitudinal low-voltage area on the posterior RA wall (median voltage, 0.05 mV). The position of the sinoatrial node is shown in Supplemental Video 2. Interestingly, the rest of the RA wall was electrically normal (median voltage, 0.35 mV) (Figure 1B).

Discussion

Giant RA is a rare congenital anomaly that causes functional tricuspid regurgitation and supraventricular arrhythmias, usually diagnosed in childhood. However, this patient had been suspected of dextrocardia since his 20s, but continued living asymptptomatically and was first diagnosed at an advanced age because of heart failure owing to supraventricular arrhythmia. There have been reports of a case of giant RA diagnosed in elderly age. Although the diagnosis

KEY TEACHING POINTS

- Acquired right atrium (RA) enlargement is common in the elderly. However, giant RA is a rare congenital disease, is usually diagnosed in childhood, and results in functional tricuspid regurgitation and supraventricular arrhythmias.
- It is extremely rare to diagnose giant RA in elderly patients, and the electrical characteristics of giant RA have not been fully elucidated.
- In this case, the posterior wall of the patient’s RA, corresponding to a part of the region originating from the sinus venous, showed a low voltage during sinus rhythm on the electrophysiology study.
- Embryonic differences, in addition to the impact of longstanding mechanical forces related to RA dilation and volume loading, may be the factors responsible for the voltage differences in a patient with a giant RA.
process of this case was similar to those of previous reports, detailed reports on the electrical features of a giant RA are scarce. Furthermore, this patient was the oldest among these reports. Several surgical reports have shown that the RA wall in such patients appeared paper-thin and translucent. Although the RA wall observed in the CT scan was paper-thin throughout, the notably thin area of the atrial myocardium on CT appears to lie laterally in this case. On the contrary, the low-voltage area was atypically limited to the posterior RA wall. The low-voltage area seems to correspond to a portion that derived from the sinus venosus, while most of the remaining RA area with preserved voltage correspond to that derived from the primitive atriums. Although the exact reason for the voltage differences between the areas is unclear, differences in embryonic origin, in addition to the impact of longstanding mechanical forces related to RA dilation and volume loading, may be the factors responsible for the voltage differences in a patient with a giant RA. In other words, embryonic differences may be responsible for the differential responses of the RA to mechanical forces related to dilation and volume loading.

Appendix
Supplementary data
Supplementary data associated with this article can be found in the online version at https://doi.org/10.1016/j.hrcr.2021.03.025.

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