Cystic tumour of the atrioventricular node: can an electrophysiological study predict sudden death?

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A 58-year-old woman presented with palpitation. A resting electrocardiogram revealed sinus rhythm and first-degree atrioventricular (AV) block (*Figure 1A*). Sporadic supraventricular/ventricular premature contractions were observed on 24 h ambulatory electrocardiogram monitoring. Transthoracic echocardiography showed a 15 mm × 17 mm round tumour in the lower interatrial septum (*Figure 1B*) that appeared as a high-density area on computed tomography (*Figure 1C*) and high and low signal intensity on T1- and T2-weighted magnetic resonance imaging, respectively (*Figure 1D* and *E*). Coronary angiography was normal. On the basis of these data, we clinically diagnosed a cystic tumour of the AV node (CTAVN). An electrophysiology study (EPS) revealed that slight prolongation of the AV interval (275 ms) and effective refractory period of the AV node (330 ms) were observed, but both were normalized with atropine. Wenckebach block occurred at 150 b.p.m., and no prolongation was observed at basal condition in any of H-V interval (40 ms) or in ventricular refractory period at either apex or outflow tract of right ventricle (240 and 220 ms, respectively). Additionally, electrical stimulations from the right ventricular apex and right ventricular outflow tract induced neither sustained ventricular tachycardia nor ventricular fibrillation. The patient underwent surgical tumour resection. The tumour was a rounded cyst filled with viscous fluid and buried in the interatrial septum in the area of the triangle of Koch. Pathological diagnosis was compatible with CTAVN (*Figure 1F*). After the cyst was completely resected, the central fibrous body was perforated with a large defect and coaptation in the tricuspid and aortic valves was severely impaired due to the loss of supportive structure. Tricuspid valve plasty and aortic valve replacement with a

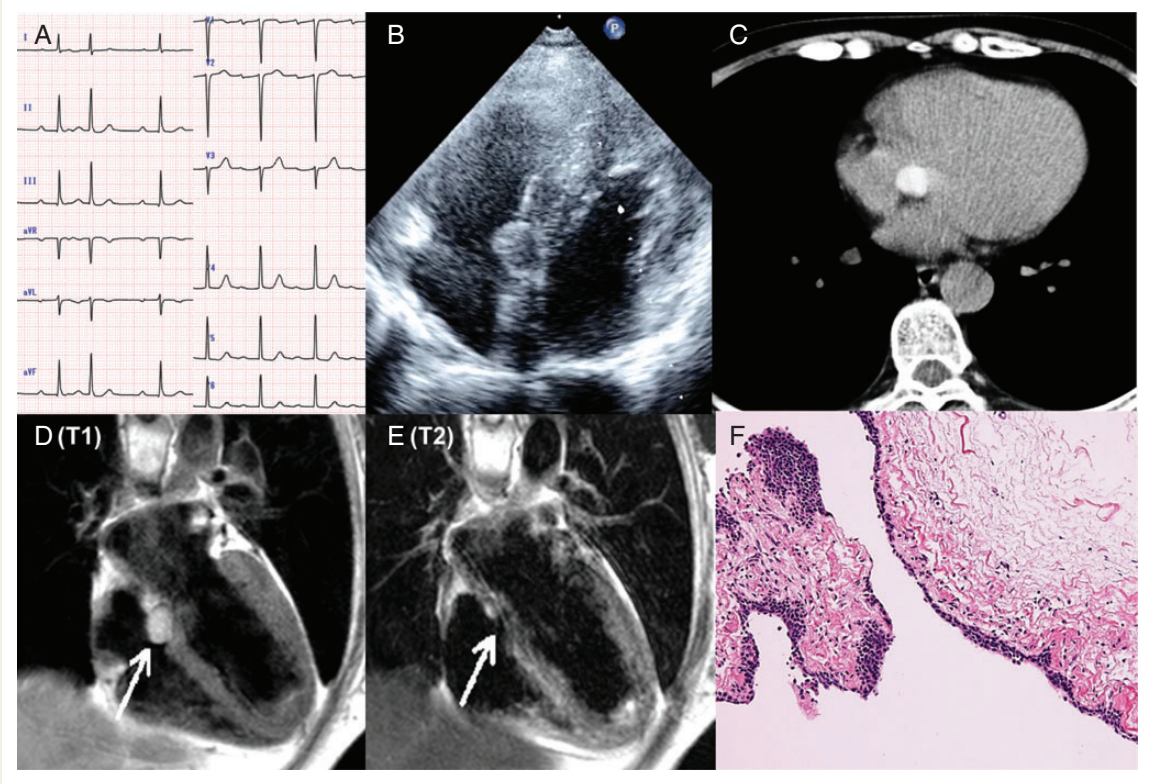

Figure 1 (*A*) Electrocardiography. (*B*) Transthoracic echocardiography. (*C*) Computed tomography. (*D*) T1- and (*E*) T2-weighted magnetic resonance imaging. (*F*) Histological staining (haematoxylin–eosin). The cyst wall was composed of fibrous connective tissue covered by stratified cells.

mechanical valve were thus performed. The patient was given a dual-chamber pacemaker post-operatively for persistent complete AV block, and no adverse event or tumour recurrence was observed during 18 months of follow-up.

Discussion

Cystic tumour of the AV node is a rare congenital cardiac tumour with a predilection for women.[1] It is located in the region of the AV node. Most cases are diagnosed incidentally post-mortem; in >1600 people who had sudden death and referred for pathological assessment, four cases (<0.25%) of CTAVN were identified.[1] Meanwhile, a few cases diagnosed ante-mortem with successful excision have been reported.[2] Cystic tumour of the AV node can cause various degrees of heart blockage and is considered the smallest tumour capable of causing sudden death[1]; however, pacemaker implantation does not prevent sudden death in these patients.[1] In some of these deaths, ventricular fibrillation was observed when patients collapsed, suggesting that CTAVN seemed to be associated with not only bradyarrhythmia but also lethal ventricular arrhythmia.[3] Surgical intervention is therefore indicated in all cases.

To our knowledge, this is the first report of CTAVN in which an EPS was performed pre-operatively. The EPS revealed only mild AV conduction disturbance responsive to atropine and no induction of sustained ventricular tachycardia/fibrillation. However, it is currently unknown whether this result indicates less or potential risk of sudden death. Further accumulation of EPS data might help to elucidate the electrophysiological cause of sudden death of CTAVN and evaluate its risk.

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