

IMAGING VIGNETTE

INTERMEDIATE

CLINICAL VIGNETTE

Cystic Tumor of the Atrioventricular Node Causing Rapid-Onset and Reversible Complete Heart Block



Alejandro Velasco, MD, Daniel J. Goldstein, MD, Leandro Slipczuk, MD, PhD, Luigi Di Biase, MD, PhD, Eric D. Manheimer, MD

ABSTRACT

Cystic tumors of the atrioventricular node are rare primary cardiac neoplasms, frequently reported as a postmortem diagnosis during autopsy. Despite their small size, they can present with complete heart block or sudden cardiac death. Herein, we present a case of rapidly progressing high-grade atrioventricular block that improved after surgery. **(Level of Difficulty: Intermediate.)** (J Am Coll Cardiol Case Rep 2023;7:101700) © 2023 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

A 33-year-old woman without cardiac history presented with palpitations, fatigue, and shortness of breath for 2 days. Electrocardiogram revealed Mobitz type 1 second-degree atrioventricular (AV) block and 2:1 AV block with narrow QRS. Electrocardiogram 3 months earlier did not show conduction abnormalities. Thyroid function and Lyme titers were unrevealing. On treadmill exercise stress test she developed complete AV block with junctional escape rhythm ([Supplemental Figure 1](#)). Transthoracic echocardiography (TTE) revealed normal left ventricular function, a thickened interatrial septum and trace pericardial effusion. Cardiac magnetic resonance imaging (CMR) showed a cystic mass infiltrating the lower interatrial septum along the AV node, with homogeneous late gadolinium enhancement ([Figures 1A and 1B](#)). A decision was made to proceed with surgical resection. A subendocardial mass was excised inside Koch's triangle that was consistent with a cystic tumor of the AV node (CTAVN) ([Figures 1C to 1F](#)). After surgery, the patient was noted to be in junctional rhythm, but later recovered AV conduction with a PR interval of 300 ms. An implantable cardioverter-defibrillator (ICD) was placed owing to high risk of permanent damage to the conduction system and a theoretical risk of sudden cardiac death (SCD). At follow-up, the patient showed normalization of AV conduction, <0.1% ventricular pacing, and no ventricular arrhythmias ([Supplemental Figure 2](#)).

From the Montefiore-Einstein Center for Heart and Vascular Care, Montefiore Medical Center, Albert Einstein College of Medicine, Bronx, New York, USA.

The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the [Author Center](#).

Manuscript received July 10, 2022; revised manuscript received November 4, 2022, accepted November 9, 2022.

**ABBREVIATIONS
AND ACRONYMS****AV** = atrioventricular**CMR** = cardiac magnetic
resonance imaging**CTAVN** = cystic tumor of the
atrioventricular node**ICD** = implantable
cardioverter-defibrillator**SCD** = sudden cardiac death**TTE** = transthoracic
echocardiography**DISCUSSION**

CTAVNs are exceedingly rare, accounting for 2.7% of primary cardiac tumors and frequently being diagnosed postmortem. Young women are most commonly affected, presenting with shortness of breath, fatigue, syncope, and SCD. High-resolution imaging with the use of CMR or cardiac computed tomography is essential, because tumors can be difficult to visualize with the use of TTE. Surgical excision is recommended to prevent further cardiac conduction involvement.

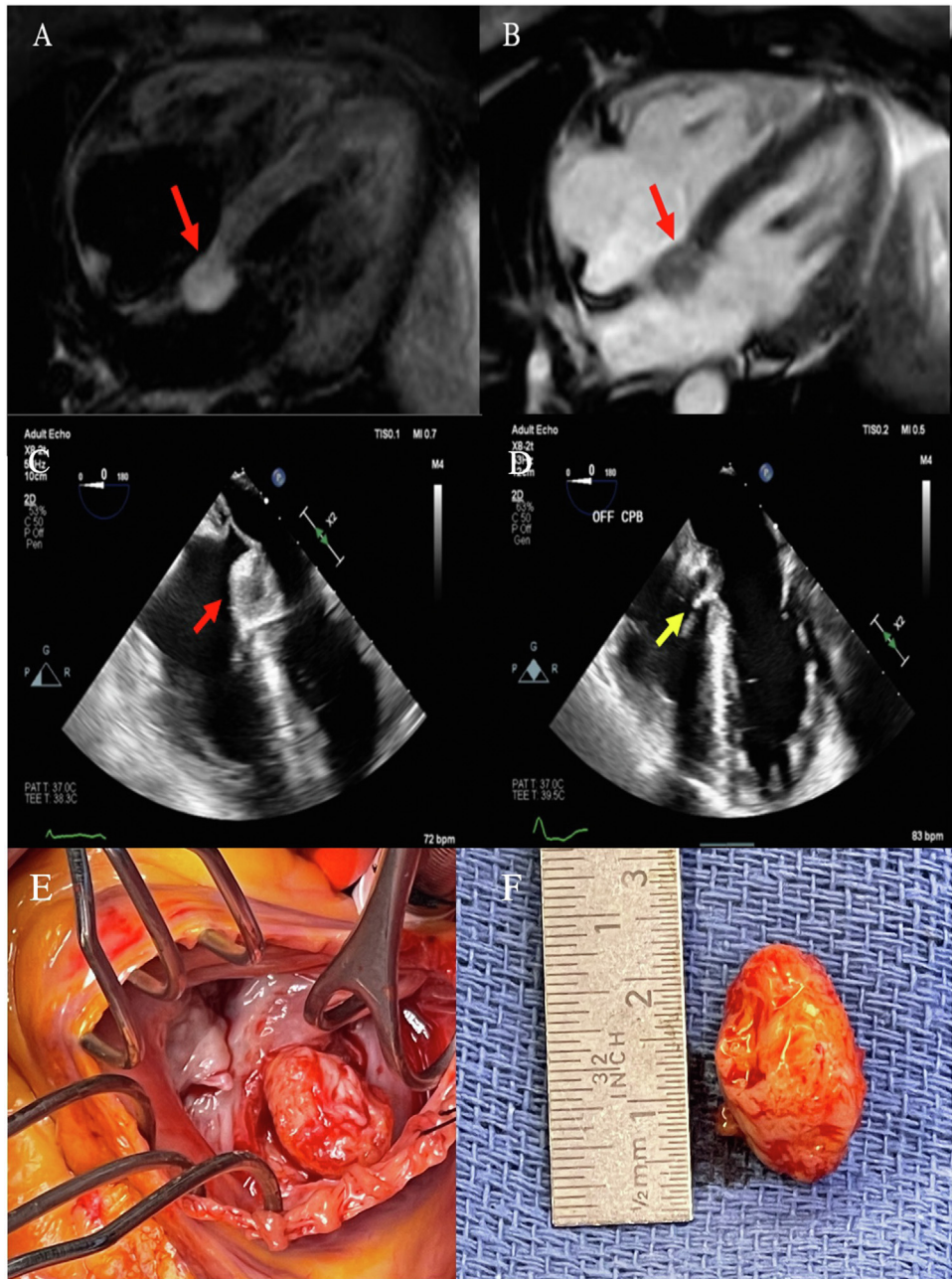
SCD in patients with CTAVN after pacemaker implantation has been reported,¹⁻³ and an ICD should be strongly considered. The mechanism for SCD and ventricular arrhythmias despite resection is unknown, but possibly related to infiltration of the ventricular myocardium/Purkinje fibers by neoplasm remnants.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

Dr di Biase is a consultant for Biosense Webster and Stereotaxis; and has received speaker honoraria/travel support from Biosense Webster, Abbott, Boston Scientific, Medtronic, Biotronik, Pfizer, and Bristol Meyers Squibb. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Eric Manheimer, Montefiore-Einstein Center for Heart and Vascular Care, Montefiore Medical Center, Albert Einstein College of Medicine, 111 East 210th Street, Bronx, New York 10467, USA. E-mail: emanheimer@gmail.com.

FIGURE 1 CTAVN



Cardiac magnetic resonance imaging of cystic tumor of the atrioventricular node (CTAVN) (red arrows) demonstrating hyperenhancement in (A) T2-weighted and (B) T1-weighted images. (C) Preoperative transesophageal echocardiographic visualization of CTAVN in the septum (red arrow). (D) Postoperative transesophageal echocardiographic image with resection and placement of pericardial bovine patch (yellow arrow) (E) Intra-operative images demonstrating CTAVN in situ. (F) Post-excision image demonstrating a spherical mass with well-defined characteristics.

REFERENCES

1. Strauss WE, Asinger RW, Hodges M. Mesothelioma of the AV node: potential utility of pacing. *Pacing Clin Electrophysiol*. 1988;11:1296-1298.
2. Patel J, Sheppard MN. Cystic tumour of the atrioventricular node: three cases of sudden death. *Int J Legal Med*. 2011;125:139-142.
3. Cohle SD. Cystic tumour of the atrioventricular node: case report and literature review. *Forensic Sci Res*. 2019;4:287-289.

KEY WORDS bradycardia, cardiac magnetic resonance, cardiac pacemaker, echocardiography

APPENDIX For supplemental figures, please see the online version of this paper.