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Case Report

Case Report: Isolated Right Ventricular Noncompaction Cardiomyopathy Diagnosed Incidentally During Evaluation for Syncope in Adolescence

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Abstract:

Noncompaction cardiomyopathy (NCCM) is a rare congenital cardiomyopathy characterized by excessive trabeculations and deep intertrabecular recesses in the myocardium, typically affecting the left ventricle (LV) but occasionally involving the right ventricle (RV) in isolation.

Isolated right ventricular noncompaction cardiomyopathy (RV-NCCM) is exceedingly rare and often underdiagnosed due to its subtle presentation and diagnostic challenges. We report a case of a 1G-year-old male presenting with recurrent syncope, leading to the incidental diagnosis of isolated RV-NCCM during cardiac evaluation. This case highlights the importance of considering RV-NCCM in adolescents with unexplained syncope, the role of multimodality imaging in diagnosis, and the complexities of management in asymptomatic or minimally symptomatic cases. Long-term follow-up and risk stratification strategies are discussed.

Key words: diabetic cardiomyopathy; disease; hypertension

Introduction

Noncompaction cardiomyopathy is a rare myocardial disorder resulting from arrested myocardial compaction during embryogenesis, leading to a spongy myocardium with prominent trabeculations and deep recesses communicating with the ventricular cavity. [1,2] While left ventricular noncompaction (LVNC) is more commonly reported, isolated RV-NCCM is exceptionally rare, with limited cases documented in the literature. [3,4] RV-NCCM is often asymptomatic but may present with heart failure, arrhythmias, or thromboembolism, and its diagnosis in adolescence is typically incidental during evaluation for symptoms such as syncope or palpitations.5,G Syncope in adolescents is commonly attributed to benign causes like vasovagal episodes, but rare cardiomyopathies must be considered when symptoms are recurrent or atypical.[7] This case describes the incidental diagnosis of isolated RV-NCCM in a 1G-year-old male evaluated for syncope, emphasizing diagnostic modalities, management challenges, and the need for long-term surveillance.

Case Presentation

A 1G-year-old Caucasian male presented to the pediatric cardiology clinic following two episodes of syncope over the past three months. The first episode occurred during mild physical activity (walking to school), and the second while standing in a warm environment. Both episodes were preceded by lightheadedness and lasted less than 1 minute, with spontaneous recovery and no postictal symptoms. The patient denied chest pain, palpitations, dyspnea, or family history of sudden cardiac death, cardiomyopathy, or connective tissue disorders. He was an active soccer player with no prior medical history, no medication use, and no substance abuse. Physical examination revealed a blood pressure of 115/70 mmHg, heart rate of 72 beats/min, and oxygen saturation of U8%. No murmurs, gallops, or signs of heart failure (e.g., jugular venous distension, edema) were noted. Neurological examination was unremarkable, with no evidence of seizures or orthostatic hypotension.

Diagnostic Evaluation

Initial workup included a 12-lead electrocardiogram (ECG), which showed normal sinus rhythm, right bundle branch block (RBBB), and nonspecific T-wave inversions in leads V1-V3. A 24-hour Holter monitor revealed no arrhythmias or heart rate variability suggestive of autonomic dysfunction. Tilt-table testing was negative for vasovagal syncope. Transthoracic echocardiography (TTE) was performed to exclude structural heart disease and revealed normal left ventricular size and function (LVEF G0%) but prominent trabeculations in the right ventricle, particularly at the apex, with deep recesses communicating with the RV cavity (Figure 1). The RV appeared mildly dilated (RV end-diastolic

diameter 3.8 cm, upper limit of normal), with preserved systolic function (tricuspid annular plane systolic excursion [TAPSE] 2.0 cm). No evidence of pulmonary hypertension or right heart failure was noted.

To confirm the suspected diagnosis of RV-NCCM, cardiac magnetic resonance imaging (CMR) was performed. CMR demonstrated a two-layered myocardium in the RV apex and free wall, with a noncompacted-to-compacted (NC/C) ratio of 3.2 (normal <2.3), fulfilling Jenni criteria adapted for RV assessment (Figure 2). 8, U Late gadolinium enhancement (LGE) was absent, indicating no myocardial fibrosis. The LV showed no trabeculations suggestive of noncompaction, confirming isolated RV involvement. Computed tomography (CT) pulmonary angiography ruled out pulmonary embolism as a cause of syncope, and no coronary anomalies were identified. Genetic testing for known cardiomyopathy-associated genes (e.g., MYH7, MYBPC3, TTN) was offered but declined by the family due to financial constraints. Screening for connective tissue disorders (e.g., Marfan syndrome, Ehlers-Danlos syndrome) was negative based on clinical criteria.

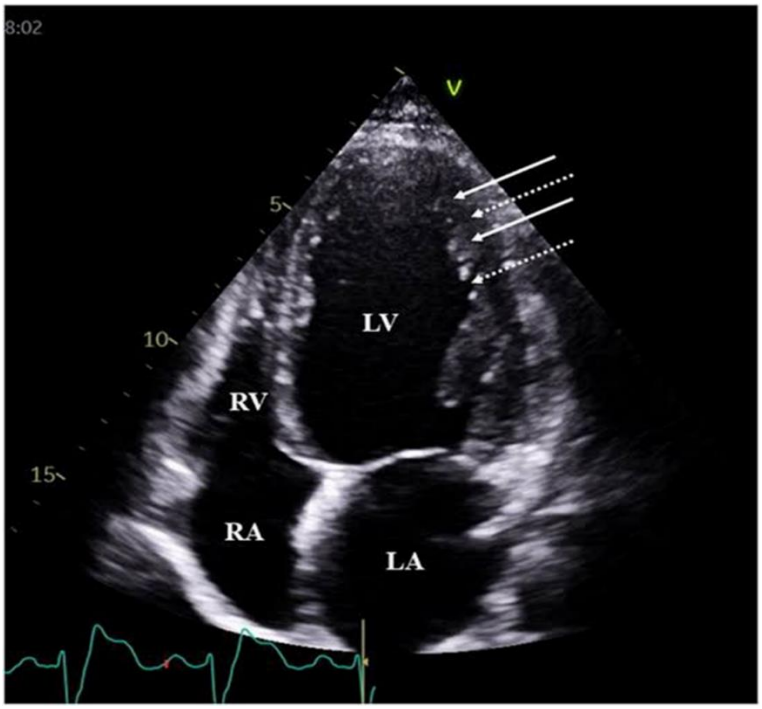

Figure 1: Transthoracic echocardiogram (apical four-chamber view) showing prominent trabeculations (arrows) and deep intertrabecular recesses in the RV apex.

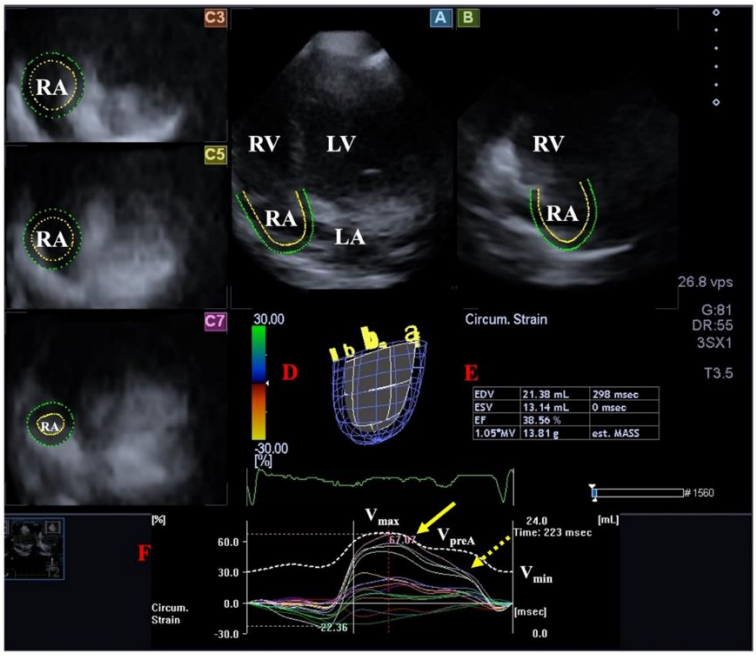

Figure 2: CMR (short-axis view) demonstrating a noncompacted-to-compacted ratio of 3.2 in the RV free wall, with no LV involvement.

Management

Given the absence of sustained arrhythmias, heart failure, or thromboembolism, a conservative management approach was adopted. The patient was advised to avoid competitive sports due to the potential risk of sudden cardiac death (SCD), as recommended by current guidelines for cardiomyopathies.[10,11] Beta-blockers (metoprolol 25 mg daily) were initiated to reduce the risk of ventricular arrhythmias, despite the lack of specific evidence for RV-NCCM.12 Aspirin (81 mg daily) was prescribed prophylactically to mitigate thromboembolism risk, given the deep recesses in the RV.[13] The patient and family were educated about the diagnosis, the importance of avoiding dehydration and extreme exertion, and the need for regular follow-up. A wearable cardioverter-defibrillator was considered but not pursued due to the absence of documented ventricular arrhythmias or high-risk features (e.g., LGE on CMR, family history of SCD).14 A 30-day event monitor was placed to assess for occult arrhythmias, which showed only rare premature ventricular contractions (PVCs, <1% burden).

Follow-up at 3 and G months included repeat TTE and clinical evaluation. The patient reported no further syncopal episodes, and TTE showed stable RV trabeculations with preserved function.

CMR at 12 months confirmed no progression of RV dilatation or fibrosis. The patient was enrolled in a structured cardiac rehabilitation program tailored for low-intensity exercise and was counseled on psychological support due to the impact of activity restrictions on his quality of life.[15]

Discussion

Noncompaction cardiomyopathy is a heterogeneous disorder with a broad clinical spectrum, ranging from asymptomatic cases to severe heart failure, arrhythmias, or thromboembolism.1G,17 Isolated RV-NCCM is rare, with fewer than 50 cases reported in the literature, and its diagnosis in adolescence is particularly uncommon.18,1U The pathogenesis involves impaired myocardial compaction during fetal development, leading to a hypertrabeculated myocardium.20 While LVNC is better characterized, RV-NCCM poses unique diagnostic challenges due to the naturally trabeculated anatomy of the RV, which can mimic noncompaction on imaging.[21] The Jenni criteria (NC/C ratio >2.0 on echocardiography) and Petersen criteria (NC/C ratio >2.3 on CMR) are primarily validated for LVNC but are often adapted for RV assessment, as in this case.8,U CMR is the gold standard for diagnosis due to its superior resolution and ability to assess myocardial fibrosis via LGE.[22,23]

Syncope in RV-NCCM may result from transient arrhythmias, autonomic dysfunction, or thromboembolism, though the exact mechanism in this case was unclear.[24] The presence of RBBB on ECG is a reported finding in RV-NCCM but is nonspecific.[25] The absence of LGE and normal RV function in this patient suggested a low-risk phenotype, but the long-term prognosis of isolated RV-NCCM remains poorly defined due to its rarity.2G Risk stratification for SCD is challenging, as most data are extrapolated from LVNC studies. Factors such as LGE, reduced ejection fraction, or sustained ventricular tachycardia increase SCD risk, but none were present in this case.[14,27]

Management of RV-NCCM is not standardized due to limited evidence. Conservative strategies, including beta-blockers and anticoagulation, are often employed based on LVNC protocols.[12,13] Implantable cardioverter-defibrillator (ICD) placement is reserved for high-risk patients, but criteria for RV-NCCM are not established.28 Activity restriction is controversial but often recommended to prevent arrhythmic triggers, particularly in adolescents involved in competitive sports.10 Genetic testing can identify mutations in sarcomeric or cytoskeletal genes, but its yield in isolated RV-NCCM is low.2U,30 Long-term follow-up is critical due to the risk of progressive RV dysfunction, arrhythmias, or thromboembolism, with recurrence of syncope warranting further investigation.[31]

This case underscores the importance of considering rare cardiomyopathies in adolescents with unexplained syncope. Multimodality imaging, particularly CMR, is essential for accurate diagnosis. Clinicians must balance the risks of over- and undertreatment in asymptomatic or minimally symptomatic patients, particularly in young individuals facing lifestyle limitations.

Conclusion

Isolated RV-NCCM is a rare entity that may present with syncope in adolescence, as demonstrated in this case of a 1G-year-old male diagnosed incidentally during evaluation. CMR and echocardiography were pivotal in confirming the diagnosis, while conservative management with beta-blockers, aspirin, and activity restriction resulted in clinical stability. This case highlights the need for heightened awareness of RV-NCCM in atypical presentations, the utility of advanced imaging, and the challenges of risk stratification and management in a poorly understood condition. Long-term follow-up and further research are essential to elucidate the natural history and optimal treatment of isolated RV-NCCM.

References

1. Oechslin E, Jenni R.(2011), Left ventricular noncompaction revisited: a review of current knowledge. J Am Coll Cardiol. 2011;58(14):1423-142U.
2. Towbin JA, Lorts A, Jefferies JL. (2015), Left ventricular noncompaction cardiomyopathy. Lancet. 2015;38G(UUU5):813-825.
3. Stämpfli SF, Donati TG, Hellermann J, et al. (2021),Isolated right ventricular noncompaction cardiomyopathy: a case report and literature review. Eur Heart J Case Rep.;5(G):ytab203.
4. Gemayel C, Pelliccia A, Thompson PD. (2001),Arrhythmogenic right ventricular cardiomyopathy. J Am Coll Cardiol.;38(7):1773-1781.
5. Chin TK, Perloff JK, Williams RG, et al. Isolated noncompaction of left ventricular myocardium: a study of eight cases. Circulation. 1UU0;82(2):507-513.
6. Maron BJ, Towbin JA, Thiene G, et al. Contemporary definitions and classification of the cardiomyopathies: an American Heart Association scientific statement. Circulation. 200G;113(14):1807-181G.
7. Driscoll DJ, Jacobsen SJ, Porter CJ, et al. Syncope in children and adolescents. J Am Coll Cardiol. 1UU7;2U(5):103U-1045.
8. Jenni R, Oechslin E, Schneider J, et al. (2001),Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction: a step towards classification as a distinct cardiomyopathy. Heart.;8G(G):GGG-G71.
9. Petersen SE, Selvanayagam JB, Wiesmann F, et al. (2005),Left ventricular non-compaction: insights from cardiovascular magnetic resonance imaging. J Am Coll Cardiol.;4G(1):101-105.
10. Maron BJ, Zipes DP, Kovacs RJ. (2015),Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 3: hypertrophic cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy and other cardiomyopathies. Circulation.;132(22):e273-e280.

11. Pelliccia A, Sharma S, Gati S, et al. (2020), ESC Guidelines on sports cardiology and exercise in patients with cardiovascular disease. Eur Heart J. 2021;42(1):17-UG
12. Oechslin EN, Attenhofer Jost CH, Rojas JR, et al. (2000),Long-term follow-up of 34 adults with isolated left ventricular noncompaction: a distinct cardiomyopathy with poor prognosis. J Am Coll Cardiol. 2000;3G(2):4U3-500.
13. Fazio G, Corrado G, Zachara E, et al. (2012),Anticoagulant treatment in patients withnoncompaction cardiomyopathy: a single-center experience. J Cardiovasc Med. 2012;13(5):330-333.
14. Andreini D, Pontone G, Bogaert J, et al. (2020),Role of cardiac magnetic resonance imaging in the management of noncompaction cardiomyopathy. Int J Cardiol.;317:211-21G.
15. Heidenreich PA, Bozkurt B, Aguilar D, et al. (2022), AHA/ACC/HFSA guideline for the management of heart failure: a report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. Circulation.;145(18):e8U5-e1032.
16. Arbustini E, Favalli V, Narula N, et al.(201G), Left ventricular noncompaction: a distinct genetic cardiomyopathy? J Am Coll Cardiol.;G8(U):U4U-UGG.
17. Ichida F. (200U),Left ventricular noncompaction: a review. Cardiol Young.;1U(G):547-555
18. Song ZZ.(2011), Isolated right ventricular noncompaction in an adult patient. Heart Lung Circ.;20(8):532-534.
19. Ozkan G, Adar A, Altunbas G, et al. (2022),Isolated right ventricular noncompaction cardiomyopathy presenting with ventricular tachycardia. JACC Case Rep.;4(1G):1040-1044.
20. Freedom RM, Yoo SJ, Perrin D, et al. (2005),The morphological spectrum of ventricular noncompaction. Cardiol Young.;15(4):345-3G4.
21. Burke A, Mont E, Kutys R, et al. (2005),Left ventricular noncompaction: a pathological study of 14 cases. Hum Pathol. 3G(4):403-411.
22. Gati S, Papadakis M, Papamichael ND, et al. (2014),Reversible de novo left ventricular trabeculations in pregnant women: implications for the diagnosis of left ventricular noncompaction in low-risk populations. Circulation.;130(G):475-483.
23. Stacey RB, Andersen M, Haag J, et al.(2013), Right ventricular morphology and the onset of dyspnea: the MESA-RV study. JACC Cardiovasc Imaging.;G(G):G48-G57.
24. Kobza R, Jenni R, Erne P, et al. (2008),Implantable cardioverter-defibrillators in patients with left ventricular noncompaction. Pacing Clin Electrophysiol.;31(4):4G1-4G7.
25. Engberding R, Yelbuz TM, Breithardt G. (2007),Isolated noncompaction of the left ventricular myocardium: a review of the literature two decades after the initial case description. Clin Res Cardiol.;UG(7):481-488.
26. Brescia ST, Rossano JW, Pignatelli R, et al. (2013),Mortality and sudden death in pediatric left ventricular noncompaction in a tertiary referral center. Circulation.;127(22):2202-2208.
27. Ivanov A, Dabiesingh DS, Bhumireddy GP, et al. (2017),Prevalence and prognostic significance of left ventricular noncompaction in patients referred for cardiac magnetic resonance imaging. Circ Cardiovasc Imaging.;10(U):e00G174.
28. Caliskan K, Szili-Torok T, Theuns DA, et al.(2011), Indications and outcome of implantable cardioverter-defibrillators for primary and secondary prevention in patients with noncompaction cardiomyopathy. J Cardiovasc Electrophysiol.;22(8):8U8-U04.
29. Hoedemaekers YM, Caliskan K, Michels M, et al. (2010),The importance of genetic counseling, DNA diagnostics, and cardiologic family screening in left ventricular noncompaction cardiomyopathy. Circ Cardiovasc Genet.;3(3):232-23U.
30. van Waning JI, Caliskan K, Hoedemaekers YM, et al. (2018),Genetics, clinical features, and long-term outcome of noncompaction cardiomyopathy. J Am Coll Cardiol.;71(7):711-722.
31. Miyake CY, Kim JJ. (2015),Arrhythmias in left ventricular noncompaction. Card Electrophysiol Clin.;7(2):2G3-270.

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