Ventricular tachycardia as the initial presentation of missed Kawasaki disease in a teenager

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Introduction

Ventricular tachycardia (VT) has been reported in acute cases of Kawasaki disease. VT secondary to ischemic sequelae is also a known long-term complication of Kawasaki disease, typically seen 2 decades after initial disease onset.[1,2] We report the first case, to our knowledge, of an otherwise healthy teenager with sustained VT as the presenting symptom for missed Kawasaki disease and describe the management approach by our team that had not considered Kawasaki disease in the differential. This case broadens our understanding of Kawasaki disease presentations in the young and timing of long-term sequelae. In light of the increasing cases of coronary dilation secondary to COVID-19 sequelae, considering ischemic mechanisms in the differential diagnosis of pediatric new-onset VT may potentially become more important in the future.

KEY TEACHING POINTS

- Ventricular tachycardia (VT) is a rare complication but can be the initial presenting symptom for untreated and missed diagnosis of Kawasaki disease even in young patients.
- When considering the differential diagnosis of new-onset VT in an otherwise healthy child, ischemic VT from acquired coronary inflammatory diseases should be considered.
- When evaluating an electrocardiogram with right bundle branch block and left axis/superior deviation, other mechanisms other than Belhassen VT should be considered, particularly when the QRS duration is >130 ms.

Case report

A 14-year-old 66 kg boy was brought to the emergency department by emergency medical services owing to chest pain, nausea, and palpitations that developed suddenly while playing video games at home. The patient was an active soccer player with no significant past medical history and had been in good health, with no recent viral symptoms or fever, prior to the onset of symptoms. He denied any prior history of cardiac symptoms such as chest pain, palpitations, shortness of breath, near syncope, or syncope. He was not taking medications and denied any over-the-counter or illicit substances. His family history was benign. Emergency medical services documented a wide complex tachycardia at a rate of 230 beats per minute (bpm) in a hemodynamically stable child. Upon arrival to the emergency department, he was found to be afebrile, tachycardic, and normotensive, with normal oxygen saturation. Physical exam showed an oriented and conversing teen with palpable pulses and evidence of good perfusion. His cardiac auscultation was only notable for tachycardia without murmurs. A 12-lead electrocardiogram showed wide complex tachycardia (Figure 1) at a rate of 230 bpm with a right bundle branch block pattern and left axis deviation (superior axis). The QRS duration was 146 ms; the VA relationship could not be definitely determined. Cardiac point-of-care ultrasound showed a grossly structurally normal heart without cardiac tumors and normal-appearing systolic function in the setting of tachycardia. He received 2 doses of adenosine (6 mg, 12 mg) with no effect.

The differential at this point included Belhassen VT using the posterior fascicle, papillary muscle VT, or other reentrant or automatic VT. Although the QRS duration was slightly longer (146 ms vs <130 ms) and the rate faster than would

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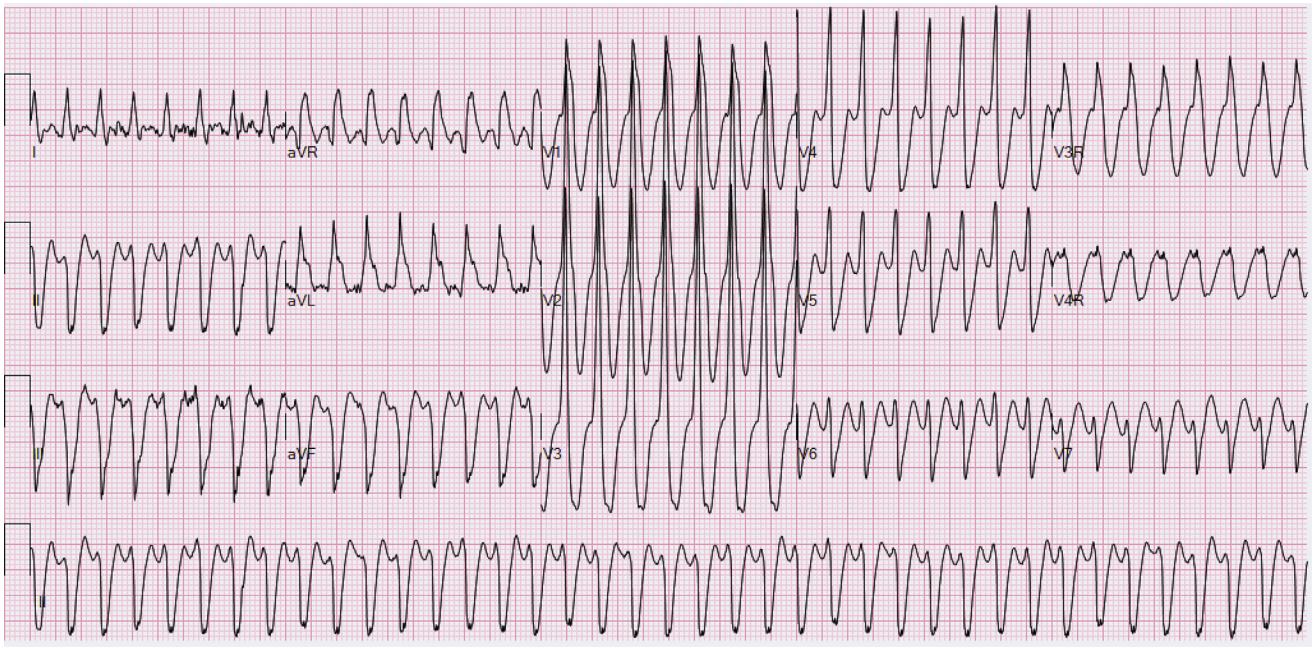

Figure 1 Electrocardiogram on arrival to the emergency department demonstrating ventricular tachycardia with a right bundle branch block pattern, left axis deviation, and superior axis. The QRS duration is 146 ms.

be expected for a fascicular VT, the decision was made to trial a calcium channel blocker. Two doses of intravenous (IV) verapamil (5 mg, 10 mg) were administered without change. He remained hemodynamically stable and comfortable. To provide the safest environment to continue further management, he was transferred to the cardiac intensive care unit. At this point his VT rate was noted to have mild fluctuations, with rates between 220 and 235 bpm. A papillary muscle VT or focal automatic VT was considered.

Given his hemodynamic stability and the possibility that DC cardioversion may not be effective if the mechanism was automatic, the decision was made to continue antiarrhythmic management. The interventional lab team was also notified in case chemical or DC cardioversion was unsuccessful. He received 2 boluses of 1 mg/kg lidocaine; the first transiently decreased the rate of the VT to 218 bpm with quick return to 230 bpm, but the second had no effect on rhythm or rate. Given the lack of response to the second bolus of lidocaine and his continued hemodynamic stability, the decision was made to trial a single IV dose of sotalol, followed by DC cardioversion attempt if sotalol was not effective. If the DC cardioversion failed, the plan was to proceed to the interventional lab. The decision was made to not administer amiodarone based on the multiple antiarrhythmics he had already received and the potential effects it may have should an electrophysiology study be pursued. He was started on an IV 1 mg/kg sotalol infusion. At our institution, an IV sotalol bolus is typically administered over 1 hour. Owing to the prior administration of verapamil and lidocaine, sotalol was ordered to be administered more slowly over 1.5 hours. However, 25 minutes after starting sotalol, the patient developed facial pallor and reported feeling dizzy and nauseated. The sotalol was immediately discontinued and an IV fluid bolus was ordered. Before receiving the fluid bolus, he became acutely hypotensive and unresponsive. He remained in VT at a rate of 230 bpm. Brief cardiopulmonary resuscitation was initiated and a 100 J synchronized cardioversion was administered, resulting in immediate conversion to sinus rhythm. He immediately regained consciousness, sat up, and reported feeling back to his normal self.

His baseline electrocardiogram demonstrated no evidence of acute ischemia or prior infarct (Figure 2). An echocardiogram was performed and revealed regional wall motion abnormalities; severely dilated left and right coronary arteries measuring 7.2 mm and 5.2 mm, respectiv and moderately depressed function (ejection fraction 37%), which quickly improved to low-normal range (ejection fraction 51%–55%) by the following morning. The appearance of the dilated coronary arteries raised immediate concerns for Kawasaki disease, although other vasculitis disorders were investigated, and a rheumatology consult was obtained. Extensive questioning for history of possible remote Kawasaki disease was negative. He had no constitutional symptoms and laboratory evaluation was not consistent with an autoimmune vasculitis. COVID-related symptoms were not considered, since the patient presented prior to the COVID pandemic. Computed tomography imaging was classic for sequelae from Kawasaki disease,[2,3] demonstrating mild diffuse right coronary artery (CA) dilation with multiple focal aneurysms; a large, calcified left main CA aneurysm impinging on the origin of the left anterior descending CA; and a diffusely hypoplastic left circumflex CA with restricted flow and flow reversal distally, suggestive of collateral flow (Figure 3). Cardiac magnetic resonance imaging demonstrated a large transmural infarct of the basal anterior and anterolateral left

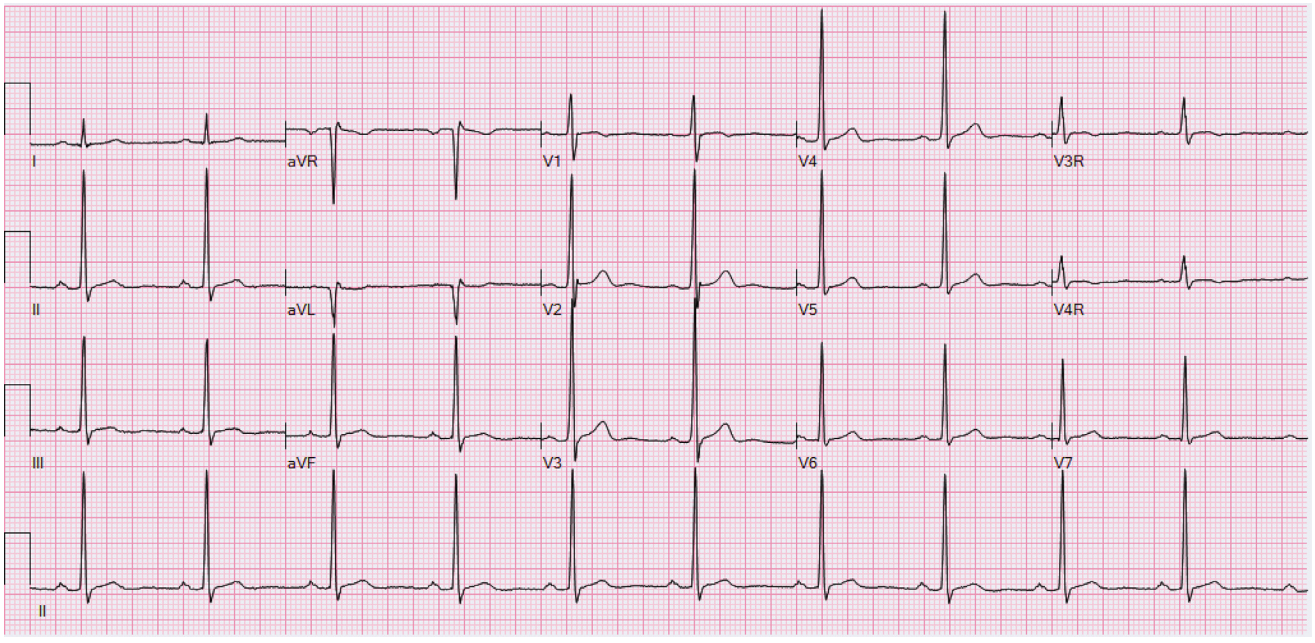

Figure 2 Baseline electrocardiogram.

ventricle. These findings were consistent with remote history of untreated Kawasaki disease.

Based on guideline criteria,[4] he was categorized as having CA disease with large and persistent CA aneurysms. He was started on warfarin, baby aspirin, and atorvastatin. His VT was felt to be most likely reentrant and secondary to scar associated with infarct. Lidocaine was administered until metoprolol was initiated the following day. He continued to have frequent ventricular ectopy and rare ventricular couplets on metoprolol (1.5 mg/kg/day) and was switched to nadolol (1.5 mg/kg/day). On nadolol, higher-grade ectopy was suppressed with the exception of occasional isolated premature ventricular contractions. He underwent an exercise stress test, which demonstrated rare isolated premature ventricular contractions without inducible VT. Lengthy discussion regarding implantable cardioverter-defibrillator (ICD) involved pediatric and adult electrophysiology teams and heart failure and Kawasaki disease experts. The risks and benefits of an ICD to prevent sudden cardiac death (SCD) were discussed with the patient and his family,[5] who ultimately declined the ICD. The decision was made to implant a looping event recorder and the patient was provided a personal automated external defibrillator. He had no further VT during his hospitalization and was discharged home on nadolol as a single-agent antiarrhythmic agent. Close follow-up with remote monitoring via his event recorder revealed a single 6-beat run of nonsustained VT at 180 bpm 6 months after discharge. The family desired medical management and nadolol was increased to 2 mg/kg/day. He has had no further events during 18 months of follow-up.

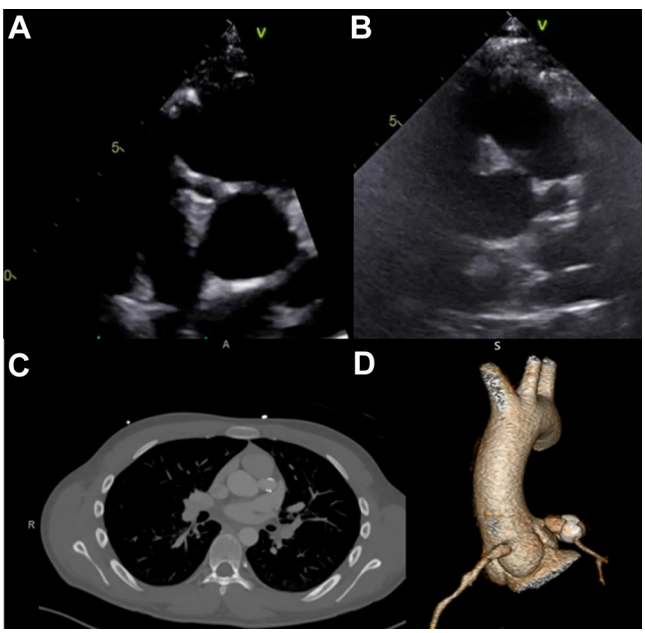

Figure 3 Imaging of coronary aneurysms. **A:** Right coronary artery with multiple fusiform aneurysms. **B:** Left coronary artery with large peripherally calcified (echo bright) aneurysm. **C:** Computed tomography (CT) angiogram of the coronary artery showing the left coronary artery with a large aneurysm with peripheral calcifications. **D:** Three-dimensional CT reconstruction of the coronary arteries showing the smaller right-sided aneurysms and the larger, calcified, left coronary artery aneurysm.

Discussion

VT that originates from ischemia-related scar, while common in adults, is less likely to be on the differential diagnosis for an otherwise healthy child presenting with new wide complex tachycardia. Although this scenario may be rare, missed Kawasaki disease does occur. Furthermore, unrecognized coronary sequelae from SARS-CoV-2 may be a contributor to late-presentation arrhythmia in the future. This case report highlights untreated Kawasaki disease as a rare cause of VT and the importance of keeping ischemia- and scar-related

mechanisms on the differential when managing new wide complex tachycardia even in pediatric populations. We provide new insights on rare late presentation of Kawasaki disease and an opportunity to learn management strategies that may be more effective in the future. In this patient, the approach followed the differential diagnosis of a wide complex tachycardia. Supraventricular tachycardia with aberrancy, fascicular or Belhassen VT, and papillary muscle VT were considered when an echocardiogram demonstrated a structurally normal heart without tumors and normal function.[6–8] Fascicular and papillary muscle VT exhibit a right bundle branch block and left/superior axis deviation, but fascicular VT tends to have a narrower QRS <130 ms. Fascicular VT has been noted to have an rR′ or rsR′ pattern in V_1 whereas papillary muscle VT is more likely to have an R or qR pattern.[7–10] Directed management for these types of VT in a stepwise manner was unsuccessful and ultimately the patient became hemodynamically unstable, resulting in the need for synchronized cardioversion to restore sinus rhythm. Once CA dilation was noted by echocardiogram, the differential diagnosis broadened to include Kawasaki disease, but perhaps should have been considered earlier in the management process.

Although Kawasaki disease has been well described for many years now, we continue to learn about atypical presentations of the disease.[11–13] When patients present in an atypical fashion, they have a higher likelihood of being missed during acute illness and not receiving timely treatment with intravenous immunoglobulin, increasing the risk of CA aneurysm by ~25%.[11] As a result, patients with CA aneurysms have a higher risk of myocardial infarction, arrhythmias, and SCD.[4,14] There have been many reports since the initial description of the disease in 1967 that describe patients with missed diagnosis of Kawasaki disease. A study by Gordon and colleagues[15] estimates that in 2009 there would have been 8400 young adults with history of Kawasaki with an AHA risk level II or greater, and this number will continue to increase yearly. This has led to an increasing interest from the adult cardiology field looking into the long-term consequences of Kawasaki disease, most importantly early coronary artery disease and SCD, among those whose aneurysms do not resolve.[15] These sequelae are more commonly being described in young adults without risks for coronary artery disease (hyperlipidemia, smoking, hypertension) who present with an acute ischemic event, most commonly 2 decades after the onset of acute disease.[1,2]

We believe it is important to share our experience in an effort to learn how to more effectively recognize and manage this scenario in the future. The initial echocardiogram in the emergency room was focused on major defects and overall function. Though we suspect the relatively rapid rate of the VT would have limited our imaging abilities, more detailed imaging of the coronaries should be considered, particularly when escalating care. With the rate of the VT and wider QRS, one could consider eliminating Belhassen from the differential. The calcium channel blocker in combination with the sotalol may have increased the risk of hypotension. We could have also considered administering more lidocaine, using amiodarone instead of sotalol, or DC cardioversion sooner. The exact approach will likely differ by physician comfort and treating center, but the overall take-home point is to consider acquired inflammatory processes (such as Kawasaki disease and perhaps even COVID-19) affecting coronary arteries as a source of VT even in asymptomatic children. We hope that details from this case can provide useful information to help direct and improve care in the future.

Conclusion

Kawasaki disease is a well-described childhood disease known to cause a vasculitis preferentially affecting the coronary arteries. Although ventricular arrhythmias are relatively uncommon, VT can be the initial presenting symptom even in the young. This case increases our knowledge of a rare late presentation of Kawasaki disease and should remind clinicians to broaden the differential diagnosis in a patient with new-onset VT, particularly when typical management approaches are unsuccessful.

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