Disabling Palpitations in an Adolescent

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A teenage girl presented with recurrent episodes of palpitations for 6 consecutive years. The resting 12-lead electrocardiogram showed sinus rhythm with no preexcitation or repolarization abnormalities. There were incessant multiple runs of non-sustained ventricular tachycardia with right bundle branch block morphologic features. QRS complexes were negative in the rest of the precordial leads (V2-V6) and in leads I and aVL, but positive in III, aVR, and aVF, suggesting a left (lateral) ventricular apical origin (Figure 1). The echocardiogram showed no structural abnormalities. The patient was prescribed sotalol but her symptoms did not improve. Invasive electrophysiologic mapping of the left ventricle endocardium demonstrated that the ventricular tachycardia probably originated from the epicardial surface of left ventricular apex.

Figure 1. Twelve-lead surface electrocardiogram during tachycardia. QRS complexes are positive in V1, negative in the rest of the precordial leads (V2-V6), and negative in leads I and aVL but positive in III, aVR, and aVF.

WHAT WOULD YOU DO NEXT?

A. Map and ablate the epicardium
B. Add a second antiarrhythmic drug and reassess the need for epicardial intervention after 3 to 6 months
C. Implant a cardioverter defibrillator
D. Perform cardiac magnetic resonance imaging to define pathologic substrate
Diagnosis
Incessant ventricular tachycardia of unknown cause

What To Do Next
D. Perform cardiac magnetic resonance imaging to define pathologic substrate

Before considering further treatment options, careful evaluation of a possible substrate is necessary. Cardiac magnetic resonance imaging is superior to echocardiography, which had normal results in this case, in detecting small tumors and myocardial fibrosis.

Discussion
Cardiac magnetic resonance imaging was requested for possible identification of a cause for the ventricular tachycardia and demonstrated a well-defined intramyocardial mass at the apical lateral wall of the left ventricle, bulging into the epicardial region. The lesion elicited low signals during the T1 and T2 black-blood sequences. Ten minutes after injection of a gadolinium-based contrast agent, the cardiac magnetic resonance image showed intense central contrast uptake with a surrounding nulled rim of myocardium, suggestive of fibroma (Figure 2A). The patient was referred to surgery where excision of a rounded 2-cm mass was performed. The tumor did not have a well-defined capsule and had to be removed by sharp dissection, including a thin layer of myocardial tissue that appeared to be mingled with the periphery of the firm tumor (Figure 2B). Histopathologic examination showed infiltrative margins of myocytes, proliferation of spindle cells, abundant network of collagen, and entrapped myocytes, confirming the diagnosis of primary cardiac fibroma (Figure 2C and D). The patient had an uneventful postoperative course and had no recurrence of the arrhythmia after the surgery.

This case emphasizes the importance of raising the suspicion of cardiac tumor as a cause of incessant tachycardia in children and young adults with otherwise normal resting electrocardiogram results. Fibroma is the second most common primary cardiac tumor in the pediatric population, after rhabdomyoma. Multiple previous reports have associated fibromas with ventricular arrhythmias and sudden death in the young. Although transthoracic echocardiography is usually the first line of imaging of cardiac tumors, it may miss small tumors owing to its limited field of view and limited resolution for soft tissue. Transesophageal echocardiography provides better views but still has limited tissue characterization abilities. Therefore, cardiac magnetic resonance imaging is considered the criterion standard for assessment of suspected cardiac tumors. Reentry is the widely acceptable mechanism of arrhythmia, as most fibromas present with monomorphic ventricular arrhythmias; however, triggered activity cannot be excluded. Ventricular tachycardia was successfully induced and entrained during electrophysiological testing in a few reported cases. In our patient, the histopathologic findings showed islands of myocardium embedded within the network of collagen and fibroblasts. This finding provides further evidence supporting reentry as the mechanism of tachycardia in cardiac fibromas. Surgical excision of the tumor was curative for the clinical arrhythmia.

Patient Outcome
During a 3-month follow-up period, the patient remained symptom free and a second electrocardiogram showed no recurrence of the ventricular tachycardia.

ARTICLE INFORMATION
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