A 14-Year Old Boy with Sore Throat and Syncope

Saira Siddiqui, MD; and Farrukh N. Jafri, MD

A 14-year-old boy presented to the emergency department with a syncopal episode. The patient stated he was walking when he suddenly became flushed and diaphoretic. He experienced a witnessed loss of consciousness of 2 to 3 minutes without seizure-like activity or a post-ictal period. He had experienced a similar episode 2 years earlier when he had become dehydrated after being physically active for a prolonged period of time; he was told it was caused by heat exhaustion. The patient denied any history of alcohol or drug use, or family history of sudden cardiac death. On review of systems, the patient stated he had a sore throat for 2 days with intermittent chills, as well as decreased oral intake for 1 day.

On examination, the patient’s vital signs included an oral temperature of 102.1°F, blood pressure of 124/69 mm Hg, heart rate of 122 beats per minute, respirations of 18 per minute, and room air oxygen saturation of 99%. The patient was a well-appearing adolescent male in no acute distress. His exam was significant for an exudative pharyngitis with anterior cervical lymphadenopathy, as well as a rapid regular heart rate with a prominent S2.

An electrocardiogram (ECG) was performed upon arrival to the emergency department (see Figure 1). After receiving 500 mg ibuprofen, 500 mg penicillin VK for streptococcus pharyngitis, and a 500-mL normal saline bolus, the patient had a repeat ECG performed (see Figure 2, page 404). Six minutes later, he had a documented oral temperature of 99.2°F. The patient was transferred to a hospital with a pediatric cardiologist. The following morning, a repeat ECG was performed (see Figure 3, page 404), and an echocardiogram found no evidence of structural cardiac abnormalities.

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For diagnosis, see page 404

Editor’s note: Each month, this department features a discussion of an unusual diagnosis. A description and images are presented, followed by the diagnosis and an explanation of how the diagnosis was determined. As always, your comments are welcome via email at Pediatrics@Healio.com.
Diagnosis:

Brugada Syndrome, Type 1

The ECGs in Figures 1 and 2 are notable for cove-shaped ST elevations larger than 2 mm in leads V1 to V3, which are followed by negative T waves in leads V1 and V2. These findings, in the absence of electrolyte or structural cardiac abnormalities, are classic signs of Brugada syndrome (BS), type 1. The ECG in Figure 3 demonstrates sinus bradycardia with resolution of the cove-shaped ST and T wave changes. These findings suggest BS unmasked by fever.

DISCUSSION

BS was first described in 1992 as a new syndrome consisting of syncopal episodes and/or sudden cardiac death in individuals with structurally normal hearts but with an ECG demonstrating ST segment elevations in leads V1 to V3 with a morphology resembling a right bundle branch block. The causes of syncope and death are attributed to prolonged QT intervals that precede acceleration of heart rate and rapid ventricular tachycardia or ventricular fibrillation that often occur without warning.1

BS has been linked to various mutations involving SCN5A, a gene encoding the alpha subunit of a cardiac sodium channel.2 It has been discovered that this mutation demonstrates arrhythmogenicity at elevated body temperatures, suggesting that patients with BS may be at higher risk during febrile states for sudden cardiac death.3

This patient’s syncopal episode and abnormal ECG warranted further exploration of possible cardiac etiologies. Long QT syndrome, hypertrophic cardiomyopathy, catecholaminergic polymorphic ventricular tachycardia, and arrhythmogenic right ventricular cardiomyopathy can present as syncope in an otherwise healthy individual. Given the morbidity associated with these conditions, they should also be ruled out in patients presenting with syncope.

There are sporadic case studies reporting dynamic changes in ECGs in febrile patients. Many of these cases with ECGs consistent with BS lack an accompanying syncopal episode and are often discovered unintentionally with resolution of the Brugada morphology upon treatment of fever; thereby forgoing further management other than routine follow-up.4-10 There are even fewer case reports demonstrating syncope in a patient with diagnosed BS requiring invasive management.11 In this unique case, the patient presented with syncope while febrile with pharyngitis and had an ECG indicating the possibility for BS. Given this patient’s history of prior
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syncope and the dynamic changes in his ECG after his fever was reduced, he was deemed to be at high risk for sudden cardiac death from BS. Per consensus guidelines, an implantable cardioverter-defibrillator (ICD) was placed and the patient discharged with pediatric cardiology follow-up. 12

CONCLUSION

Although BS is an extremely rare disease, a pediatric patient with a syncopal episode warrants evaluation with ECG, particularly in the context of febrile illness. The patient’s underlying illness should be evaluated and fever treated expeditiously. Suspicious ECGs should be evaluated by a pediatric cardiologist, with ICD placement considered for patients presenting with syncope and ECG changes consistent with BS. ■

REFERENCES