Advanced Emergency Nursing Journal
Vol. 38, No. 2, pp. 109–114

CASES OF NOTE

Column Editor: Kathleen Sanders Jordan, DNP, RN, MS, FNP-C, SANE-P

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A Zebra Among Horses

A Case of Brugada Syndrome and Coronary Artery Disease

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ABSTRACT
Brugada syndrome (BrS) is recognized as a hereditary ion channel disorder with electrocardiographic changes. First appearing in the literature about 20 years ago, contemporary thoughts are that BrS may be responsible for many sudden cardiac deaths and is associated with ventricular dysrhythmias that can lead to syncope or cardiac arrest. Many individuals with BrS may have no or limited structural heart disease, whereas others may have subtle morphological changes in histopathology. This case reviews a single patient with BrS Type 1 who was found to have a high-level of coronary artery disease. Changes noted in the original electrocardiogram were of significant importance in reaching the diagnosis. **Key words:** Brugada syndrome, CAD, coronary artery disease, ion channel disorder

PATIENTS who experience syncopal events often present to emergency departments (EDs) for evaluation and treatment. Cardiovascular examinations and electrocardiographic (ECG) findings are included, along with a neurological examination, as part of the initial assessment for many patients. Timely recognition, intervention, and documentation of baseline findings are critical for patients who experience cardiovascular disease. This case describes a middle-aged patient who experienced a syncopal event and was subsequently transported to the ED by emergency medical services (EMS) for further evaluation. The emergency physician noticed critical findings in the electrocardiogram that revealed Brugada syndrome (BrS) Type 1 in the presence of high-level coronary artery disease (CAD) during the initial assessment; such findings warranted interventional cardiology and hospital admission. Emergency departments are commonly the first points of contact for patients with cardiovascular emergencies; therefore, it is critical for clinicians to timely recognize and treat CAD and BrS Type 1.

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Disclosure: *The authors report no conflicts of interest.*

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DOI: 10.1097/TME.0000000000000102

CASE REPORT

A 51-year-old man presented to the ED for evaluation of a syncopal event that he experienced while at a shopping mall with his wife in the afternoon. The patient was briefly unconscious, which led to initiating the community EMS. While en route to the ED, an infield electrocardiogram by EMS suggested ST-segment elevation myocardial infarction (STEMI); therefore, the paramedics activated the cardiovascular catheterization laboratory (Cath Lab) protocol. The patient arrived at the ED, and a second electrocardiogram revealed BrS Type 1. His initial vital signs were a blood pressure of 113/72 mmHg, heart rate of 82 beats per minute, respirations of 20 breaths per minute, and temperature of 37.06°C, and he denied pain. His oxygen saturation was 93%.

Chief Complaint

The patient reported intermittent shortness of breath that began after a syncopal event, diaphoresis, nausea, and lightheadedness.

History of Present Illness

Prior to the syncopal event, the patient was not experiencing any distress. The syncopal event had an abrupt onset with loss of consciousness that was witnessed by his family members.

Medical, Social, and Familial History

The patient reported a history of hypertension and seasonal allergies and reported that his hypertension and seasonal allergies were controlled with lisinopril and Zyrtec (cetirizine). He denied any previous surgical history. He reported that he does not smoke tobacco and that he rarely consumed alcoholic beverages. He denied previous syncopal events, cardiac dysrhythmias, and CAD. The patient denied a familial history of premature CAD, dysrhythmic events, or cardiac surgical procedures.

Initial Physical Examination and Review of Systems

Upon initial physical examination, the patient was 51-year-old man who was well groomed and well nourished. He illustrated no signs of acute distress. He presented with moderate diaphoresis. The patient was alert and oriented to self, place, time, and situation, with all cranial nerves intact and functioning within defined limits. His HEENTM (head-ears-eyes-neck-throat-mouth) examination revealed normal inspection, and he denied trauma and pain. His respiratory system revealed clear breath sounds bilaterally with no respiratory distress. He reported intermittent shortness of breath. His cardiovascular system revealed a regular rate and rhythm. He was negative for edema, bruits, thrills, jugular venous distention, and murmur. The patient's abdomen was nontender and soft. He reported nausea and recent episode of diarrhea. His back revealed normal inspection and was negative for vertebral point tenderness and costovertebral tenderness. The patient's extremities had a range of motion within defined limits, with no evidence of injury or edema. The patient's skin revealed a normal color for race and was warm and dry without alterations in skin integrity. He was negative for lymphadenopathy.

Course of ED Treatment

The patient underwent an initial physical examination, chest radiography, 12-lead ECG studies, and laboratory studies including complete blood cell count, comprehensive metabolic profile, brain naturietic peptide (BNP), coagulation studies, and point-of-care (POC) troponin. The patient was prophylactically given 2 L of oxygen, and he was monitored continuously with a cardiac monitor and pulse oximetry. Automated blood pressure readings were obtained periodically. The electrocardiogram in the ED revealed ST-segment elevation in the precordial leads (V_1–V_3) (see Figure 1). The initial laboratory findings were unremarkable, and acute myocardial infarction was ruled out.

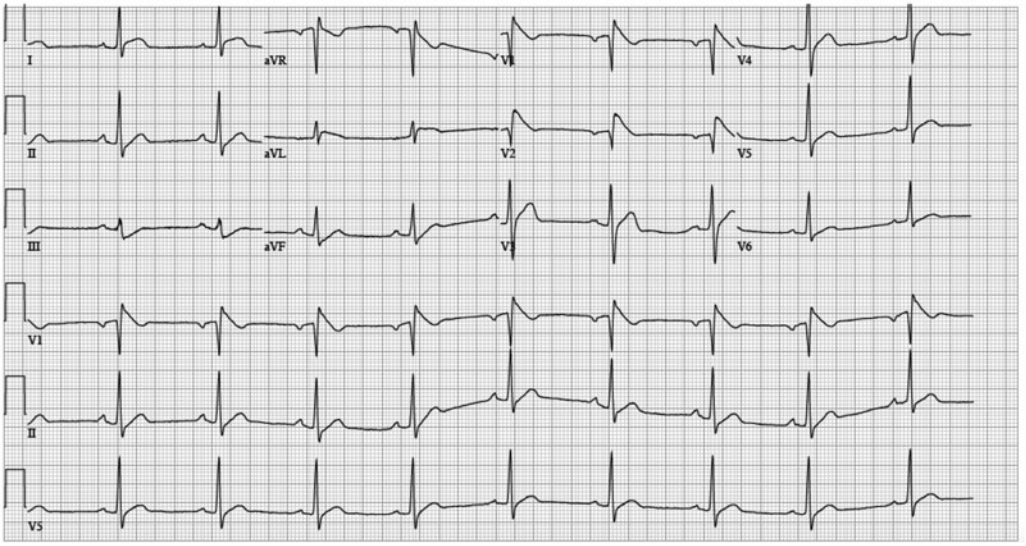

Figure 1. Patient's presenting ECG tracing.

The patient's POC troponin was less than 0.04 ng/ml and BNP was less than 100 pg/ml. The chest radiograph revealed clear lungs without effusions and a normal hear and mediastinum (see Figure 1).

Cardiology Consult

The patient's initial electrocardiogram was concerning for STEMI; therefore, the emergency physician consulted with interventional cardiology. The interventional cardiologist requested administration of Brilinta (ticagrelor) 180 mg and aspirin 325 mg orally. Closer inspection of the patient's electrocardiogram suggested BrS Type 1, thus warranting consultation with electrophysiological cardiology. The electrophysiologist confirmed the suspicion of BrS Type 1 due to the pseudo-right bundle branch block and persistent ST-segment elevation in leads V_1–V_3 and recommended that the interventional cardiologist proceed with cardiac catheterization.

Assessment/Diagnosis/Plan of Care

On the basis of the ECG findings, the patient was diagnosed with an acute STEMI and BrS Type 1. The patient required immediate cardiac catheterization and admission to the intensive care unit (ICU).

Cardiac Cath Lab

The patient was taken to the Cath Lab and was found to have severe multivessel CAD with 90% occlusion that required surgical interventional (e.g., cardiac bypass). The patient did not undergo angioplasty or receive stents at that time. He was immediately taken to the ICU.

Post-Cardiac Catheterization Outcomes

Once in the ICU, he received an echocardiogram that revealed an ejection fraction of 69%–70%. The patient received a consultation with electrophysiology, and the electrophysiologist recommended an internal defibrillator after the patient's CAD was under control. He underwent a three-vessel coronary bypass surgery with saphenous graft and 7 days later he underwent internal cardiac defibrillator implantation. The patient was discharged home to his family after 11 days in the hospital.

DISCUSSION

Clinicians first reported BrS a little more than 20 years ago (Brugada, Campuzano, Sarquella-Brugada, Brugada, & Brugada, 2014). Brugada syndrome held distinct ECG patterns, primarily patterns of right bundle branch block

accompanied by ST-wave elevation in the right precordial leads (V_1–V_3). The syndrome was also associated with a high incidence of sudden cardiac death (SCD), especially in younger-than-expected adult populations. Ventricular tachyarrhythmia, ventricular fibrillation, and SCD were also noted in such patients who also had structurally normal hearts (Arbelo & Brugada, 2014; Brugada et al., 2014; Gray, Semsarian, & Sy, 2014).

Presently, BrS is classified as a familial channelopathy that most often involves the inward sodium current (de Luna, Garia-Niebla, & Baranchuk, 2014; Gray et al., 2014). The SCN5A gene (Brugada et al., 2014) is responsible in up to 75% of cases (Arbelo & Brugada, 2014). More than 100 other genes and electrolyte channels also contribute to the disorder. The inherited pattern is one of autosomal dominance with variable penetrance (de Luna et al., 2014), although the pattern may be absent in up to 60% of other relatives (Arbelo & Brugada, 2014). Identification of gene mutation alone is insufficient for diagnosis (Arbelo & Brugada, 2014).

Epidemiology/Incidence

The true worldwide incidence of BrS in the general population is unknown (Arbelo & Brugada, 2014). There are current estimates, however, across regions of the world. In the United States and Europe, the incidence is suggested to range between 1 and 5 per 10,000 individuals. A somewhat higher incidence is noted for Asia, with reports ranging between 5 per 1,000 and 12 per 10,000 being suggested (Arbelo & Brugada, 2014; Franco, Dias, Teresa, & Hebert, 2014). Prevalence in males occur eight to 10 times more so than in females, which has been attributed to differences in the transmembrane ionic current differences between the genders and attributed to higher testosterone levels (Arbelo & Brugada, 2014; Mashar, Kwok, Pinder, & Sabir, 2014). Likewise, prognosis differs by gender, with males having up to a 5.5-fold increase in the risk for SCD. Also, the syndrome appears to be absent in prepubescent individuals, again, attributed to levels of testosterone. Symptoms are least experienced during periods of physical exertion. Rather, symptoms occur more frequently during nocturnal time (midnight to 6 a.m.), a variation thought to be due to variations between the sympathetic and parasympathetic nervous systems (Arbelo & Brugada, 2014).

The Electrocardiogram

In patients who have confirmed BrS Type 1 pattern, there is a characterization of a *coved type* wave appearance, meaning that there is an ST-segment elevation of 2-mm or more in more than one right precordial lead (typically V_1–V_2 but may extend into V_3), followed by T-wave inversion (Arbelo & Brugada, 2014; de Luna et al., 2014; Sheikh & Ranjan, 2014). The highest point of the ST segment must reach at least 2 mm higher than the isoelectric line in the first precordial lead (de Luna et al., 2014). There is a mismatch in QRS width between V_1 and V_6 (Sheikh & Ranjan, 2014).

In patients with BrS Type 2 pattern, *saddleback* wave morphology is noted in V_1 and V_2 with ST-segment elevation of 2 mm or more, followed by a sustained elevation of 0.5 mm above the isoelectric line and either an upright T wave or a biphasic T wave (de Luna et al., 2014; Sheikh & Ranjan, 2014). There is a mismatch in QRS width between V_1 and V_6 (Sheikh & Ranjan, 2014). See Figure 2.

ECG Electrode Placement

Specificity and sensitivity of ECG tracings in patients with BrS are variable (Holst et al., 2012). This variation can be partially explained through electrode (lead) placement. Placement of the precordial leads in higher positions (e.g., second or third intercostal space) increases the sensitivity in some BrS Type 1 patients; therefore, if diagnosis is suspected, the clinician should try higher lead placement to confirm suspicion (Arbelo & Brugada, 2014; Holst et al., 2012). Holst et al. conversely found that higher electrode placement lowered specificity for BrS Type 2 patients. However, Harrigan, Chan, and

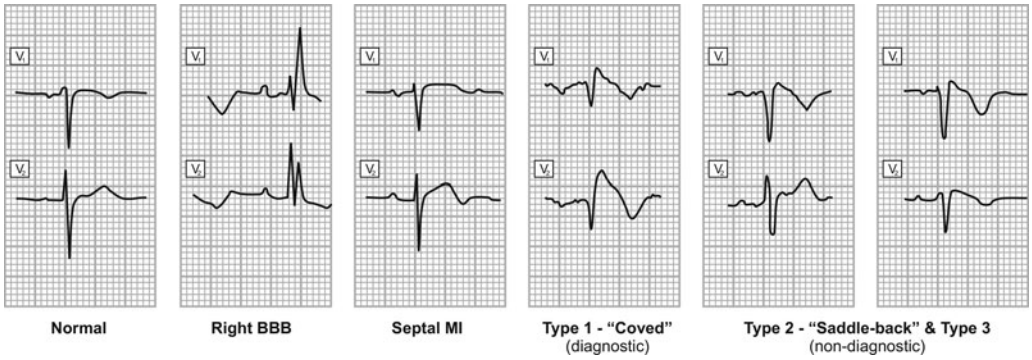

Figure 2. Comparison of wave morphology between normal, bundle branch block, and Brugada syndrome. BBB = bundle branch block; MI = myocardial infarction.

Table 1. Differential diagnoses

Isolated right bundle branch block
Arrhythmogenic right ventricular dysplasia
Acute pericarditis
Acute myocardial ischema/infarction
Pulmonary embolism
Dissection aortic aneurysm
Hyperkalemia
Hypercalcemia
Hypothermia
Duchenne muscular dystrophy
Heterocyclic antidepressant overdose
Long QT-syndrome
Friedreich's ataxia
Mediastinal tumor

Note. From de Luna et al. (2014), Nishizaki, Yamawake, Sakurada, and Hiraoka (2013), and Sheikh and Ranjan (2014).

Brady (2012) report that improper placement of the electrodes may mimic a variety of pattern changes that falsely indicate underlying pathology.

Differential Diagnoses

See Table 1 for differential diagnoses.

Management of BrS

Currently, given the risk of SCD, an implantable cardioverter defibrillator is the most effective and safe strategy for preventing SCD by terminating life-threatening dysrhythmias (Brugada et al., 2014). There are no first-line pharmacological agents suitable for consideration in treating BrS (Sheikh & Ranjan, 2014), although quinidine and isoproterenol have been explored (Mashar et al., 2014). Radiofrequency ablation has demonstrated benefit in prevention of ventricular fibrillation (Brugada et al., 2014); however, the procedure itself is not without risk (Mashar et al., 2014).

Nursing Implications

This case highlights several important aspects for professional nursing and advanced practice nursing. The initial presentation of syncope requires investigation to determine etiology. Upon registration to an ED, patients must receive a thorough cardiac, neurological, and metabolic evaluation. For patients who have cardiac conditions, having prompt laboratory, radiology, and Cath Lab personnel available is essential, given national standards in cardiac care. Emergency department registered nurses should have a solid ECG knowledge on both the recognition of bundle branch block and STEMI. For these patients, ED registered nurses should assess a familial history for cardiac dysrhythmias for patients who present with syncope and ventricular tacky rhythms.

One prudent reminder is that when reviewing an electrocardiogram with unexpected changes, the option to repeat the tracing is simple, noninvasive, and of little financial impact. The correct 12-lead

electrode placement, while seemingly minor, is of utmost importance. Having the prompt ability to consult with cardiology cannot be overemphasized when considering all the factors that contribute to ECG abnormalities. Registered nurses should stay current with advanced ECG interpretation skills, especially with 12-lead electrocardiograms.

CONCLUSION

Brugada syndrome Type 1 is truly a zebra among horses. Many individuals are unaware of the condition, and an unexplained syncopal episode may be the first indication of a potentially life-threatening condition. Having a structurally normal heart without coronary vessel disease is common.

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