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CASE REPORTS

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Transient Brugada phenocopy during evolving ischemic right bundle branch block

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ABSTRACT

A "Brugada phenocopy" (BrP) is a clinical presentation exhibiting the same electrocardiographic characteristics of true Brugada syndrome (BrS) in different clinical scenarios. We report the clinical case of a 59-year-old male patient with acute chest pain depicting BrP followed by extensive convex ST-segment elevation due to proximal thrombosis of the left descending coronary artery. Following successful percutaneous intervention and drug-eluting stent implantation, a classical right bundle branch block pattern emerged. We discussed possible mechanism and clinical implications of this unusual electrical sequence during acute myocardial injury.

Key Words: Brugada phenocopy, Infarction, Right bundle block

1. INTRODUCTION

The development of a Brugada ECG pattern (type 1 or 2) during the context of acute coronary ischemia can be due to ischemia unmasking a true Brugada syndrome (BrS) or, alternatively, an ischemic Brugada Phenocopy (BrP) in patients without persistent sodium channel dysfunction.

BrS presents a congenital physiological substrate predisposing to malignant ventricular tachyarrhythmias and sudden cardiac death.[1] Its hallmark electrocardiographic (EKG) pattern is an elevation of ST-segment through V1-V3 chest leads. However, these distinct EKG features are dynamic and can be often concealed. Attempting to unify the nomenclature under a unique scientific term, BrP is being applied to describe these cases presenting identical EKG pattern to BrS but caused by diverse pathological states; diverse clinical scenarios where an environmental condition can potentially imitate a genetic disease.[2] A systematic diagnostic approach is very useful to make a prompt and correct differentiation between true BrS and BrP (see Table 1).[2, 3]

Table 1. Current criteria to diagnose BrP

Number	Diagnosis criteria	Observed result
I	EKG pattern at baseline	Type-2 Brugada morphology
II	Identifiable underlying condition	Acute myocardial injury
III	EKG pattern evolution	Resolved after reversion of underlying condition
IV	Pretest probability of BrS based on clinical criteria	Low
V	Sodium channel blocker challenge (ajmaline, flecainide, or procainamide)*	Not done
VI	Genetic test#	Not done

* Not mandatory if surgical right ventricular outflow tract manipulation was performed within last 3 days.
# Desirable, not mandatory. [2, 3]

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The presence of a new bundle branch block is a marker of poor prognosis during acute myocardial infarction (AMI).[4] However, the prognostic impact of a new right bundle branch block (RBBB) developed during AMI remains poorly explored. We describe a patient with anterior AMI who develops a type-2 BrP evolving to classic RBBB pattern during revascularization process.

2. CLINICAL PRESENTATION AND EKG DESCRIPTION

A 59-year-old Caucasian smoker male, presented at the emergency department (ED) with chest pain at rest that appeared at 03:00 PM. The pain was dull, intense (7/10), radiated to his jaw and resolved after administering IV nytroglicerin. Standard 12-lead EKG depicted a saddle-back (type-2) Brugada pattern[3] (see Figure 1). He received a clopidogrel loading dose, aspirin, intravenous heparin and atorvastatin and was emergently transferred to the cath lab. A thrombosis occluding proximal left anterior descending coronary artery was found in coronary angiography. Then, a drug-eluting stent implantation resulted in subsequent TIMI 3 flow (see Figure 2). After percutaneous intervention, the type-2 Brugada pattern evolved to classic complete RBBB with persistent convex ST-segment elevation through V1-V6, DI-aVL. Five hours later, the EKG showed new q-waves in leads V1-V4 and persistent ST-segment elevation localized to middle chest leads (see Figure 1). The patient was discharged on enalapril, atenolol, atorvastatin, aspirin and clopidogrel 7 days after admission.
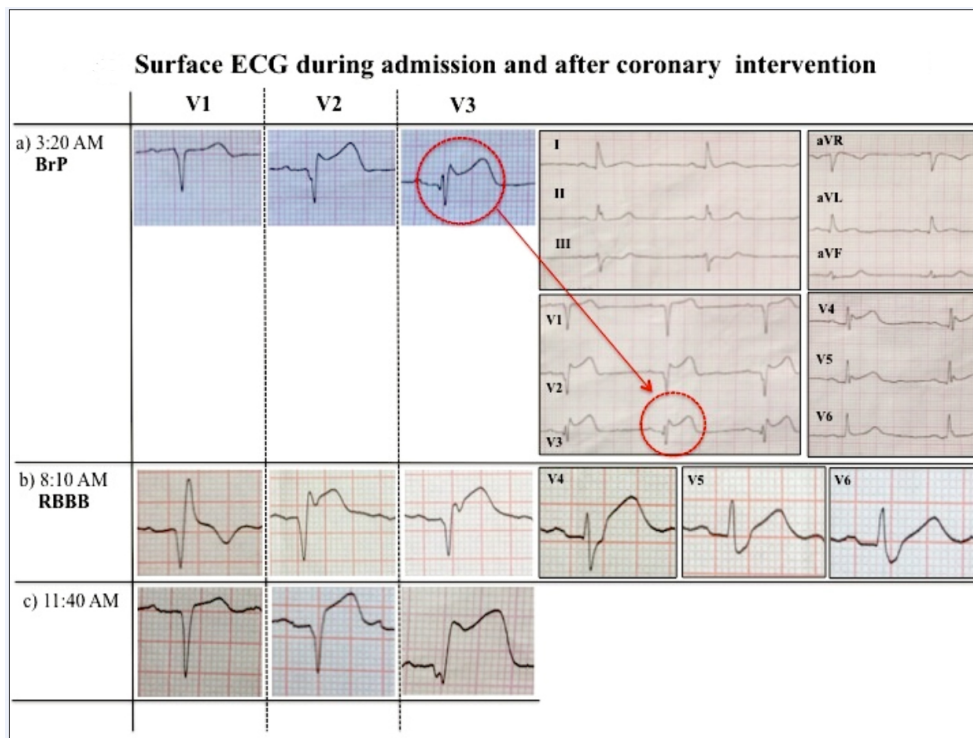

Figure 1. In-hospital surface EKG. Electrocardiographic tracings obtained during presentation (a) and on admission to the Coronary Unit (b, c). Note the snapshot of BrP morphology (a) evolving to transient RBBB immediately after percutaneous intervention (b) and to anterior Q wave 5 hours later (c)

3. DISCUSSION

A thrombotic occlusion of proximal left anterior descending coronary artery tend to cause a RBBB pattern associated with an extensive ischemic burden, since first septal perforating branch frequently supplies both the Hisian right bundle branch and the left anterior fascicle.[5] However, some diagnostic limitations arise during acute myocardial injury due to RBBB pattern. First, RBBB may potentially mask subtle anterior ST-segment elevation due to secondary depression of ST-segment after rSR deflection, and falsely depressing the ST-segment below the threshold to diagnose an acute transmural myocardial injury. As we described, another diagnostic challenge could be derived from the differential diagnosis with BrS. To confirm or discard the development of a new classical RBBB pattern, both limitations should be minimized obtaining serial EKG tracings during clinical evolution.
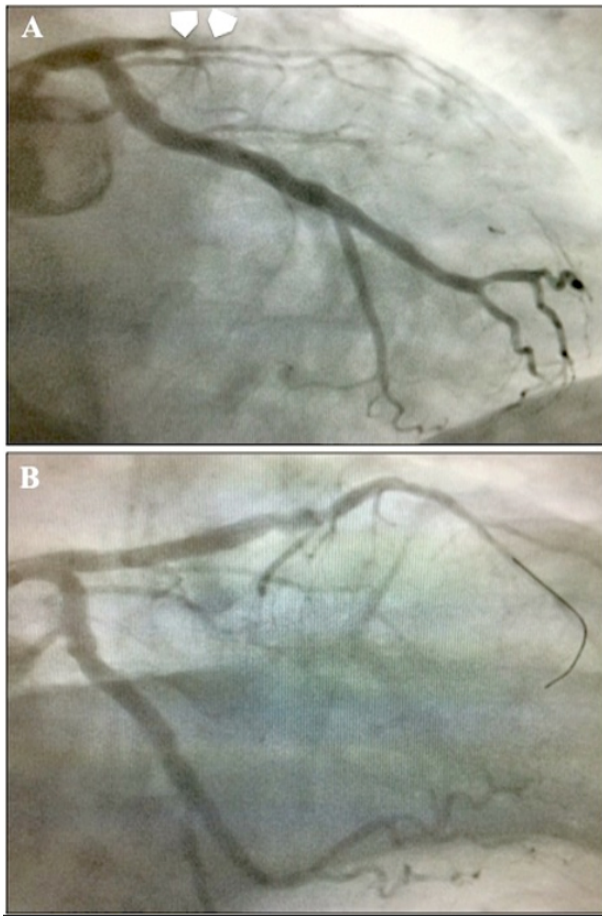
Figure 2. Coronary angiography and percutaneous intervention. White arrows in (A) showed that left anterior descendent coronary artery was proximally occluded. (B) Angiographic result after successful everolimus-eluting stent implantation (Xscience™ 3.5 mm × 23 mm).

An early and correct differentiation between BrP and true BrS is of paramount importance in this clinical setting to avoid unnecessary tests as well as not delaying coronary reperfusion. This is a really challenging task because ischemia may either induce a BrP or, conversely, unmask a BrS.[2] Based on current criteria, we confirmed a Type-2, Class B BrP in this clinical presentation.[3] However, a pharmacological challenge with flecainide or ajmalin was not performed to discard a true BrS, since sodium channel blocker may cause lethal ventricular arrhythmias during ongoing myocardial injury in patients not affected with BrS. Additionally, as SCN5A mutation is identifiable in no more than 30% of probands affected

by BrS, a genetic testing was not mandatory to diagnose BrP (see Table 1).

We found only a few clinical cases of BrP associated with acute ischemia in the literature. Tomcsányi et al. reported the clinical presentation of a 59-year-old man who presented saddled-back ST-segment elevation pattern extending from V1 to V3 and changing to coved morphology after 3 hours.[6] Another report described a 53-year-old man carrying a left anterior descending artery bridging combining EKG abnormalities of BrP or early repolarization pattern.[7] Anselm et al. referred to a 70-year-old hypertensive and diabetic male suffering an acute inferior infarction with right ventricular involvement and typical coved type-1 Brugada pattern located to V1 resolving after streptokinase administration.[8] More recently, we described a woman suffering AMI due to mid left anterior coronary occlusion evolving to classic Wellens' pattern.[9] These rare clinical presentations shows us that some BrP may be confused with an AMI and that the reverse situation can also occur.

True mechanisms underlying BrP in acute myocardial ischemia are not fully understood. A significant decrease in fast sodium current into the myocite, commonly observed in true congenital BrS, determines an imbalance between the inflow and outflow of positive charges at the end of phase 1 action potential. This altered ion channel activity generates a notch and loss in the dome of the action potential, which is due to altered outward potassium current (Ito). As the Ito density is higher in epicardial layers than in endocardium, this electrical phenomenon occurs heterogeneously through the ventricular wall. When changes in the notch are sufficiently marked, epicardial action potential is significantly longer compared to endocardium, and ST-segment elevation and T-wave negativity become evident in surface EKG. Some findings obtained in experimental BrS models have shown that Ito could modulate the EKG manifestations of myocardial ischemia and BrS, and suggested both pathological states could share an electrophysiological substrate.[11] The International Registry and the Educational Portal on BrP (www.brugadaphenocopy.com) represent an extremely useful initiative to define the epidemiology, pathophysiology and clinical evolution of these patients.

CONFLICTS OF INTEREST DISCLOSURE
The authors have declared no conflicts of interest.

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