what's your diagnosis?

Diagnosis: Brugada Syndrome

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Ann Saudi Med 2007; 27(3): 223-224

We describe a 29-year-old man who presented to our emergency department (ED) with chest pain and features of Brugada syndrome on ECG, but the diagnosis was only made after his third visit to the ED. It is important to shed the light on this rare but important syndrome since it affects young people and when diagnosed, treatment options to prevent sudden cardiac death are highly successful.

Brugada syndrome is one of the causes of sudden cardiac death in the young with no structural heart abnormalities. The current prevalence is estimated to be 0.4% to 0.6% in western countries, but a higher frequency

may be found in Far East countries like Thailand.^{1,2} The syndrome has a characteristic pattern on resting ECG, but some times a provocative test needs to be done to demonstrate the abnormality.

First described by Pedro and Joseph Brugada in 1992, Brugada syndrome is an autosomally dominant inherited cardiac disease with variable expression, resulting from mutation in the SCN5A gene, causing life-threatening polymorphic ventricular arrhythmia in a patient who is otherwise healthy. Men are more commonly affected than women. Sudden death happens usually while sleeping and is the presenting symptom of one third of patients, and is not related to exercise.³ The

Figure 2. Precordial leads of patients with Brugada syndrome showing all three patterns. From Wilde, AA Eur Heart J 2002; 23: 1648, with permission.

major pathophysiologic mechanism is loss of sodium channel function.

Brugada syndrome has a characteristic ECG pattern in the form of incomplete RBBB and ST segment elevation in the right precordial leads (V1 to V3), thought to be due to an alteration in the action potential in the epicardial but not the endocardial cells. The ECG changes may be dynamic and have transient normalization. Three different patterns of ST elevation have been described; in the classic Brugada ECG (type 1), the elevated ST segment (≥ 2 mm) descends with an upward convexity to an inverted T wave (Figure 1). Type 2 and 3 patterns have a "saddle back" ST-T wave configuration in which the elevated ST segment descends toward the baseline, then rises again to an upright or biphasic T wave. The terminal part of the ST is elevated ≥ 1 mm

in type 2 and < 1 mm in type 3 (Figure 2).^{3,4}

Diagnosis of Brugada syndrome should be suspected in patients with ST segment elevation in more than one right precordial lead (V1-V3) and one of the following:⁴ syncope, a family history of sudden cardiac death at an age younger than 45 years, typical ST segment changes in family members, nocturnal agonal breathing, documented ventricular fibrillation, or electrophysiological inducibility. The appearance of typical ECG changes alone in the absence of one of the above mentioned criteria is considered to represent an idiopathic Brugada ECG pattern but not the Brugada syndrome. In those with normal or transient ECG changes, the characteristic changes can be provoked by administration of sodium channel blockers, like flecainide or procainamide.³

REFERENCES

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