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Electrocardiogram Quiz – Case 25

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A 31-year-old man with an unremarkable past medical history, presented to the emergency department with palpitations of a few days' duration. The patient was hemodynamically stable. The 12-lead surface electrocardiogram (ECG) is depicted in figure 1.

Questions

a. What abnormalities are depicted on the 12-lead ECG (fig. 1)?
b. What is the clinical significance of the depicted abnormalities?

Comment

The Brugada syndrome (BS) is a malignant, genetically-determined, arrhythmic syndrome, manifesting as syncope or sudden cardiac death (SCD) in individuals with structurally normal hearts. The BS was first described in 1992, and since then, it had an exponential rise in the numbers of cases reported, to such an extent, that the second consensus conference reported in 2005 named it as the second leading cause of death in males <40 years (after trauma). Its incidence seems to be particularly high in Southeast Asia where it had been previously described as sudden unexplained nocturnal death syndrome (SUNDS). In the Philippines it was colloquially known as bangungut ("to rise and moan in sleep"), in Japan as pokkuri ("sudden and unexpectedly ceased phenomena") and in Thailand as Lai Tai ("death during sleep"). The mean age of sudden death is 41, with the age at diagnosis ranging from 2 days to 84 years.

The BS is often referred to as a sodium channelopathy. Over 60 different causative mutations have been described so far, and at least 50% are spontaneous mutations, but familial clustering and autosomal dominant inheritance has been demonstrated. ECG changes can be transient with Brugada syndrome and can also be unmasked or augmented by multiple factors such as fever, ischemia, hypokalemia, hypothermia, and a variety of drugs (flecainide, propafenone, calcium channel blockers, nitrates, alcohol, cocaine, etc.).

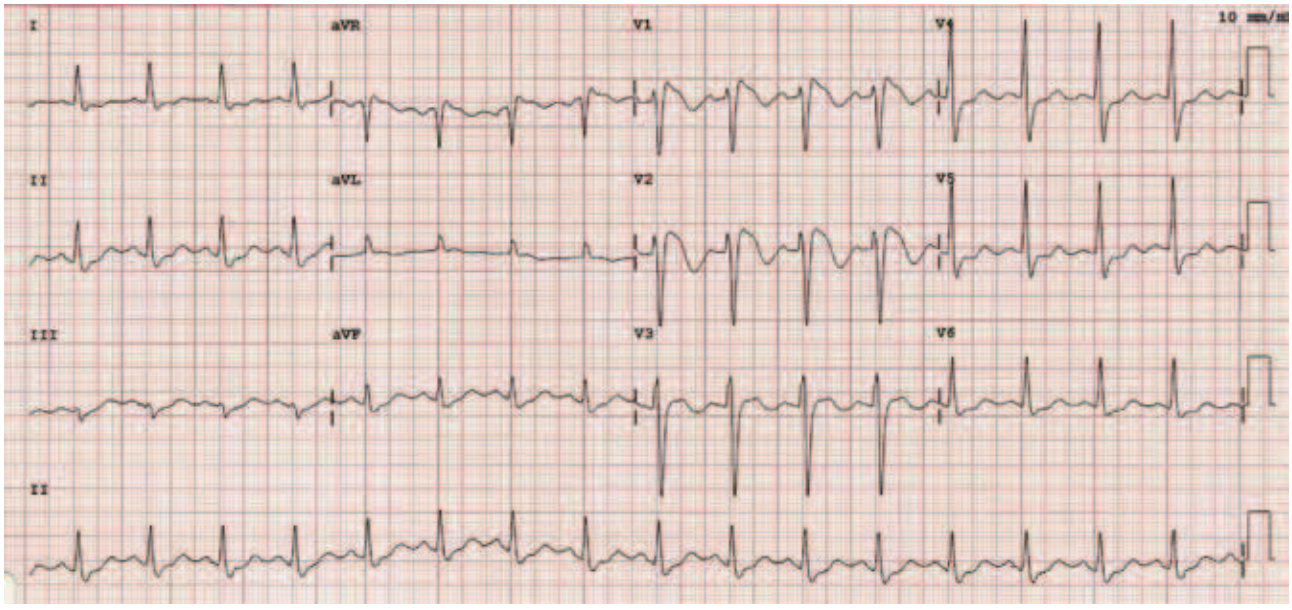

Figure 1

The symptoms associated with the BS are due to re-entry ventricular arrhythmias, typically arising in the affected zone of the right ventricle. If the arrhythmia's duration is short and terminates spontaneously, it can be asymptomatic or cause palpitations, as in our patient, syncope or nocturnal agonal respiration, or can degenerate into ventricular fibrillation (VF) and lead to cardiac arrest.

Type 1 ECG pattern (coved ST segment elevation >2 mm in >1 of V1-V3 followed by a negative T wave) is the only ECG abnormality that is potentially diagnostic, as in our patient. This has been referred to as the Brugada sign. This ECG abnormality must be associated with one of the following clinical criteria to make the diagnosis: Documented VF or polymorphic ventricular tachycardia (VT), family history of SCD at <45 years old, coved-type ECGs in family members, inducibility of VT with programmed electrical stimulation, syncope, nocturnal agonal respiration. The other two types of Brugada are non-diagnostic but possibly warrant further investigation. Type 2 pattern has a >2 mm of saddleback shaped ST elevation. Finally, Brugada type 3 can be the morphology of either type 1 or type 2, but with <2 mm of ST segment elevation.

The only proven therapy is an implantable cardioverter-defibrillator (ICD). Quinidine has been proposed as an alternative in settings where ICD's are unavailable or where they would be inappropriate (e.g. neonates). Our patient was referred to the Electrophysiology Department for further investigation and treatment.

References

1. DE LUNA AB, GARCIA-NIEBLA J, BARANCHUK A. New electrocardiographic features in Brugada syndrome. *Curr Cardiol Rev* 2014, 10:175–180
2. BAYÉS DE LUNA A, BRUGADA J, BARANCHUK A, BORGGREFE M, BREITHARDT G, GOLDWASSER D ET AL. Current electrocardiographic criteria for diagnosis of Brugada pattern: A consensus report. *J Electrocardiol* 2012, 45:433–442
3. BRUGADA P, BRUGADA J, ROY D. Brugada syndrome 1992–2012: 20 years of scientific excitement, and more. *Eur Heart J* 2013, 34:3610–3615

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