Reminder of important clinical lesson

The dilemma of an incidental preoperative electrocardiogram showing a Brugada phenotype

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A Brugada pattern on routine electrocardiography is one of several features that can indicate the potential for life threatening rhythm disturbances. The authors describe such a scenario in an asymptomatic 38-year-old woman who required significant surgery under anaesthesia. The diagnosis and possible management routes are outlined with an emphasis on the incumbent psychosocial and familial issues that are encountered. The patient underwent her surgery and was then further investigated to 'prove' the diagnosis. Ultimately, the patient declined further investigations and interventions. This case highlights the dilemma faced by anaesthetists and clinicians, not to mention the patients and their family when a Brugada phenotype is identified on a routine ECG. Clinicians should seek an expert opinion but ultimately, as in this case, the patient should be positioned to make an informed decision on what route to follow.

BACKGROUND

This case highlights the problems with the discovery of a condition that may have life changing effects on the patients and their family. This is particularly problematic when the patient is young, fully active, asymptomatic and in this case, of both childbearing age and requiring a significant surgical procedure. We have attempted to describe

the process through which one progresses when this electrocardiographic feature is uncovered.

CASE PRESENTATION

A 38-year-old Caucasian woman attended a preoperative assessment clinic prior to spinal surgery. An ECG revealed features characteristic of type 2 Brugada phenotype (figure 1A).

Figure 1 (A) Precordial leads V1-V3 of the preoperative ECG showing sinus rhythm with a ventricular rate of 57 beats per min, significant first degree heart block (per rectum (PR) interval 268 ms) and a right bundle branch block pattern. In leads V1 and V2, there is a slewed concave ST-segment elevation of 0.3 mV with T-wave inversion, compatible with a type 2 pattern Brugada phenotype. (B) Precordial leads V1-V3 after 4 min of an ajmaline challenge test (total of 28 mg administered), now showing a type 1 Brugada pattern.

Figure 2 Schematic diagram illustrating the diagnostic pathway, management and consequences. A spontaneous type 1 pattern or drug induced type 1 pattern carries the most significance for future events.^{1*} This may include postmortem cardiogenetic analysis of family members experiencing sudden death.^{2*} Aborted cardiac event includes aborted VT/VF or sudden cardiac death.^{3*} An ajmaline challenge requires counselling as to the attendant risks of inducing VT or VF.^{4*} Gene testing requires the involvement of genetic counselling.^{5*} A negative gene screen does not exclude a diagnosis of Brugada syndrome.^{6*} Patients may or may not take up the offer of an automatic implantable cardio-defibrillator.^{7*} EPS, electrophysiological cardiac studies.

She was an active equestrian and was also taking no regular medication. A previous appendicectomy had been performed under general anaesthesia without incident. The only relevant personal history was of two vasovagal episodes following routine venesection, and two episodes of short-lasting palpitations while anxious.

The family history included the unexpected cardiac death of her mother at age 63 and her maternal grandfather at age 43, the latter after developing pneumonia. In both cases the deaths were ascribed to 'heart attacks', with no further details available.

INVESTIGATIONS

The patient was offered an ajmaline challenge to further substantiate the diagnosis. The ECG demonstrated a type 1 Brugada pattern within 1 min of starting this diagnostic test and was terminated at 4 min (total dose of 28 mg of ajmaline) due to the extent of electrocardiographic change (figure 1B).

Following further full and informed discussion (written information provided) as to the benefits, risks and implications of further testing to the patient, and her family, the patient decided not to pursue further investigations or treatment.

TREATMENT

Treatments of any type or further investigations have been declined by the patient after being fully informed as to all the possibilities available.

OUTCOME AND FOLLOW-UP

The spinal surgery was conducted successfully under general anaesthesia using propofol.

DISCUSSION

Brugada syndrome is an autosomal dominant inherited defect characterised by coved ST-segment elevation of at least 2 mm in two right precordial leads with a negative T-wave (type 1 pattern). This may be spontaneous or induced by a sodium channel blocker. Two other ECG patterns are described: type 2 with a 'saddleback' ST-segment with high takeoff ST elevation of at least 2 mm gradually descending to a positive or biphasic T-wave; and type 3 with either a 'saddleback' or 'coved' appearance with ST elevation < 1 mm. Type 2 and type 3 ECGs are not diagnostic of Brugada syndrome unless they change to a type 1 pattern after administration of a sodium channel blocker such as ajmaline.

Brugada syndrome is frequently inherited as an autosomal dominant trait with variable expression and incomplete penetrance. Approximately 20% of cases are caused by mutations in the gene encoding the A subunit of the cardiac Na⁺ channel SCN5A. Other rarer genes include the glycerol-3-phosphate dehydrogenase 1-like protein; A1 and B subunits of the L-type Ca²⁺ channel (CACNA1C and CACNB2b); KCNE3 and SCN1B, which encodes the function-modifying sodium channel β-1 subunit (<1%).

Table 1 Drugs considered to be contraindicated in Brugada syndrome

Drugs to be avoided	Drugs preferably avoided	Potentially arrhythmic
Ajmaline*	Amiodarone [‡]	Isoprenaline/ isoproterenol*
Flecainide*	Disopyramide [‡]	Quinidine*
Pilsicainide*	Lidocaine [‡]	Orciprenaline [‡]
Procainamide*	Propranolol [‡]	Cilostazol [‡]
Propafenone [‡]	Verapamil [‡]	Amitriptyline [‡] / Clomipramine [‡]
Bupivacaine [‡]	Ketamine [‡]	Lithium [‡]
Propofol*	Nicorandil [§]	
Acetylcholine [‡]	Nifedipine [§] / Diltiazem [§]	
Ergonovine [‡]	Nitroglycerine [§] / Sorbidnitrate [§]	
Cocaine [‡]	Dosulepine [‡] / Fluoxetine [‡]	
	Phenytoin [‡]	
	Alcohol excess [‡]	

*Convincing evidence. [‡]Evidence less clear. [‡]Conflicting evidence. [§]Very little evidence.

Patients may present with palpitations, syncope or cardiac arrest but are frequently asymptomatic. Arrhythmias typically occur at rest or during sleep. Precipitating factors for arrhythmia include hypokalaemia, hypothermia, bradycardia, fever, myocardial ischaemia and some drugs (table 1). Table 1 is an abbreviated list based on that compiled by Postema *et al.* (2009).¹

Diagnosis rests on the demonstration of a type 1 ECG with one or more of the following features: documented ventricular fibrillation (VF) or polymorphic ventricular tachycardia (VT); family history of sudden cardiac death < 45 years old; coved ECGs in family members; inducible VT with programmed electrical stimulation; and syncope or nocturnal agonal respiration.

Our patient had experienced palpitations and developed a type 1 ECG following an ajmaline challenge. The family history was only mildly indicative of arrhythmic deaths. As frequently described, the pathognomonic ECG features were transient and, until drug-challenged, were of the type 2 phenotype. This type 2 pattern was in association with marked first degree atrioventricular block, a feature recognised to be associated with sodium channel mutations.² When suspected, confirmation of Brugada syndrome, through electrophysiology studies and/or an ajmaline challenge is usually offered.

The use of drug testing in this context is still debated. The power of these tests to uncover patients at risk of sudden cardiac death is controversial and the tests themselves carry an associated risk of provoking fatal cardiac arrhythmias.¹ However, recent data suggest that the risk of a cardiac event in an asymptomatic patient is low at 0.5% per year;³ asymptomatic subjects require no specific treatment, except for advice on avoidance of recognised

precipitant drugs, excess alcohol, stimulants and high temperatures or fever.

The patient we describe would likely not have been offered an automatic implantable cardioverter, but ideally would have been offered a baseline of annual review with Holter ECG recordings. The patient was also instructed to contact our service, should any symptoms develop, that might be indicative of rhythm disturbance. The appropriate duration of follow-up remains unknown. To date, the largest study of patients with Brugada syndrome (FINGER Brugada Syndrome Registry) concludes that even this study, with the longest published follow-up, is too short to draw final conclusions.³

A Brugada ECG alone is not definitive in diagnosing Brugada syndrome.⁴ Conducting further testing and screening holds many implications for the patient and their family. Some individuals may subsequently regret undertaking screening (including gene testing) because of implications for insurance, occupations, recreation, sport and travel. A positive test result may also have significant psychosocial consequences on the patient and their immediate family members (figure 2). However, this is a condition whereby prior knowledge may reduce the risk of sudden death where precautions are advised and followed.

Learning points

- ▶ The Brugada syndrome is a dilemma for the patients and their family.
- ▶ A potential Brugada syndrome is a dilemma for anaesthetists and clinicians.
- ▶ Clinicians should seek an expert opinion.
- ▶ Ultimately, as in this case, the patient should be positioned to make an informed decision on what route to follow.

Competing interests None.

Patient consent Obtained.

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