21. Focal Takotsubo Syndrome/ Stress Cardiomyopathy as an Initial Presentation of a Concealed Congenital Long QT Syndrome

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Body

Background: Takotsubo syndrome (TTS) or Stress Cardiomyopathy is a reversible cause of myocardial injury presenting as heart failure with regional left ventricular dysfunction after a preceding emotionally or physically stressful event. Prolongation of the QT interval in classic TTS may be observed and is typically attributed to an acquired Long QT Syndrome (LQTS). We report the first case of a Focal TTS with a normal baseline QT interval complicated by ventricular arrhythmia and diagnosed to have a concealed Congenital Long QT Syndrome 1 using the epinephrine QT stress test.

Case: A 50-year old, Filipino female consulted with a two-month history of intermittent palpitations and exertional dyspnea attributed to increasing emotional stress due to instability in finances. There was no chest pain, paroxysmal nocturnal dyspnea, orthopnea, fever and other systemic symptoms. Three days prior, the patient had a significantly heightened state of stress due to family problems and started to have bouts of non-rotatory dizziness with near syncope. A few hours prior, the patient had recurrent bouts of syncope associated with palpitations and vague chest heaviness. There was no seizure, post-ictal confusion or neurologic deficits.

At presentation she had a blood pressure of 100/70 mmHg, heart rate of 65 beats/minute, respiratory rate of 18 breaths/minute, temperature of 36.6 degrees Celsius. She had another bout of syncope associated with polymorphic ventricular tachycardia (VT) which spontaneously terminated.

Baseline electrocardiogram was sinus rhythm, normal axis with non-specific T wave inversion at V1-V2, normal corrected QT interval. The patient had low serum potassium at 3.2 mmol/dL and serum magnesium at 0.82 mmol/dL. Serial highly sensitive troponin I was normal at presentation: 12.0 ng/mL and after 3 hours: 16.9 ng/mL (NV<29ng/mL). CRP was normal at <5.0 mg/L while ESR was slightly elevated at 32 mm/hour (NV 0-20 mm/hour).

Transthoracic echocardiography revealed eccentric left ventricular hypertrophy with hypokinesia consistent with the left anterior descending artery distribution (LAD) with a depressed ejection fraction (EF) of 48%. Coronary angiography revealed angiographically normal coronary arteries.

Cardiac Magnetic Resonance Imaging (MRI) done days later revealed a focal area of thinned myocardium with mild hypokinesia at the apical cap (segment 17) suspicious for delayed transmural enhancement. Chamber dimensions were now normal with a preserved overall systolic function and an Ejection of Fraction of 61%.

Discharge impression was Focal Takotsubo Syndrome complicated by ventricular arrhythmia. Left ventricular function improved and no QT prolongation nor significant ST-T wave changes were initially documented. The patient was sent home on Nebivolol.

However, the patient was readmitted due to pulseless arrest from ventricular fibrillation. After prompt treatment and stabilization, three-dimensional echocardiography revealed left ventricular EF of 65% with no dyssynchrony nor wall motion abnormality.

Provocative testing with epinephrine QT stress test was then performed. There was a paradoxical prolongation in the QTc > 35 milliseconds at the 2nd minute which was maintained throughout the steady state phase (3-5) minutes. (Table 1.) Frequent premature ventricular depolarizations with R on T phenomenon were seen at the 4th minute with no induction of torsades des pointes or other arrhythmias.

These findings were consistent with Congenital Long QT Syndrome Type 1. The patient underwent placement of a dual chamber implantable cardioverter defibrillator (ICD) and was discharged on Propranolol and Amlodipine. On follow-up and interrogation, the ICD was able to deliver appropriate shocks for bouts of ventricular arrhythmia. This was managed by increasing the pacing rate and titrating the beta-blockade.

Discussion: Our case was a classic presentation of TTS with a focal LAD distribution wall motion abnormality and depressed systolic function. Absence of coronary artery disease, myocarditis and the documented recovery of EF further confirms this diagnosis. However, the cluster of ventricular arrhythmias was inconsistent and made us suspect an arrhythmogenic underlying cause. This prompted us to proceed with provocative testing. In the absence of genetic testing especially in low-resource settings, the epinephrine QT stress test developed by Shimuzu and colleagues was a valuable specific and sensitive test to diagnose our patient with Congenital Long QT 1 Syndrome as revealed by the pathognomonic response of paradoxical QT interval prolongation during infusion.

Our scenario demonstrated for the first time the possibility of focal TTS being the initial presentation of a concealed congenital LQT1 syndrome. We hypothesize that in the setting of physical and/or emotional stress, the surge of neuronal activation and catecholamines occurring in TTS may also be the central mechanism for the paradoxical prolongation of the QT interval to manifest.

It is possible that Congenital Long QTS Type 1 may in itself be a risk factor for TTS as its sequelae/complication. Further studies are necessary to establish a clear relationship between these two entities.

Pharmacologic therapy for the prevention of sudden cardiac death and ventricular arrhythmia remains limited for our case for which we preferred the application of an ICD as an intervention especially for our patient with documented life-threatening ventricular arrhythmia.

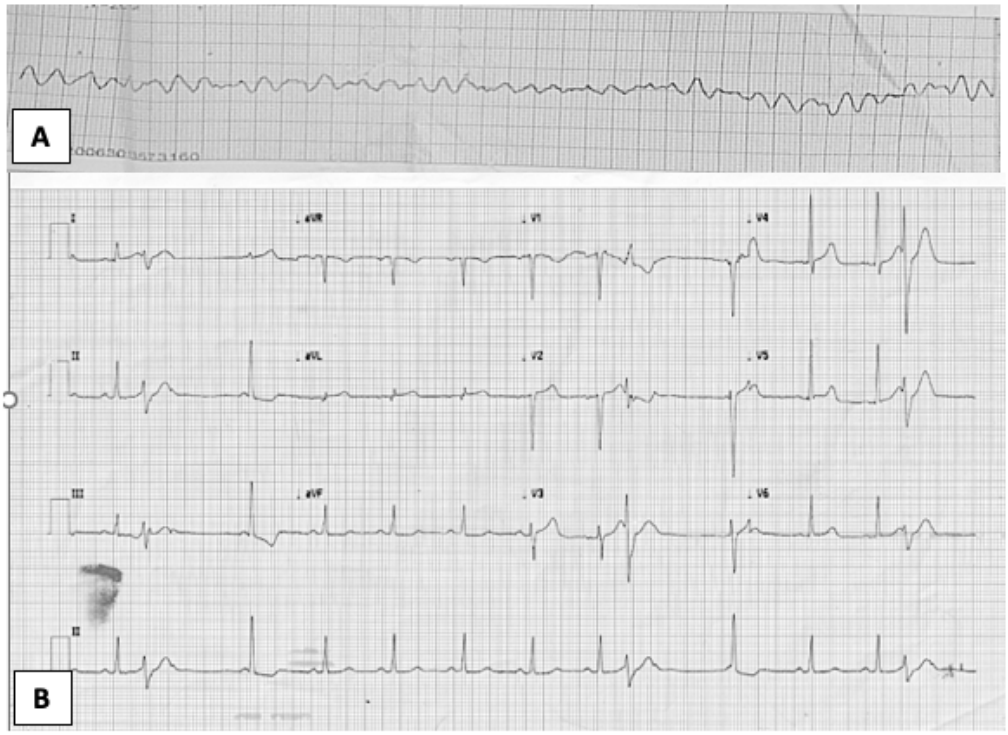

Figure 1. A. Ventricular Fibrillation B. 12 Lead Electrocardiogram post defibrillation showing sinus rhythm, normal axis, premature ventricular contractions, intermittent atrioventricular block, normal QT interval at 400 milliseconds, with R on T phenomenon.

Time	HR	Mean QTe	Mean QTp	Mean QTce	Mean QTpe	Mean QTce - QTpe	Change in QTC
Baseline	98	353	287	452	366	85	0
0:30	141	303	233	465	358	107	13
1:00	139	312	237	474	360	114	22
1:30	133	323	237	481	352	129	29
2:00	125	352	232	508	334	173	56
2:30	115	372	227	515	314	201	63
3:00	114	378	237	521	326	195	69
3:30	103	382	237	500	310	190	48
4:00	103	373	255	489	334	155	37
4:30	107	367	264	490	352	138	38
5:00	100	350	255	452	329	123	0
7:00	99	340	255	437	328	105	-15
20:00	100	342	256	441	331	110	-11

Table 1. QT intervals measured during the infusion of epinephrine using the Shimuzu protocol showing an increase in QTc at the 2nd minute that persisted throughout the steady state.