EP CASE REPORT

Hypothyroidism and congenital long QT: additive effect causing torsades?

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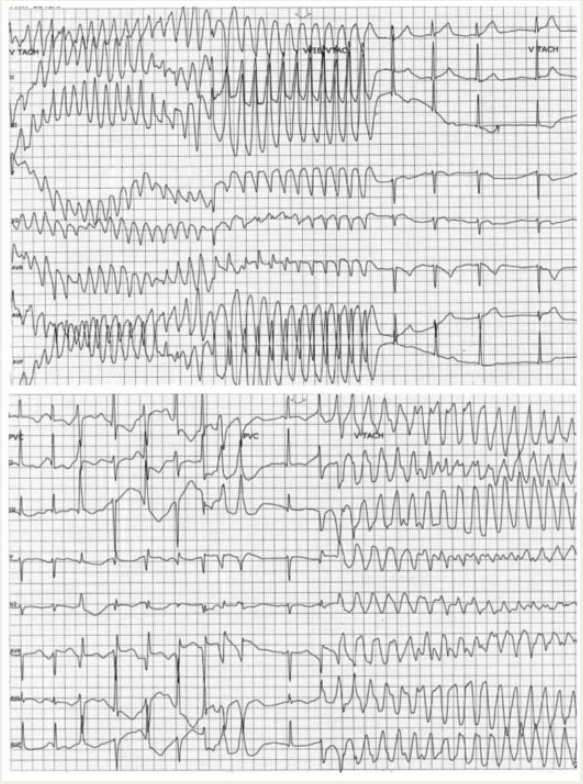
A 19-year-old woman was hospitalized following a syncope episode. Initial investigations in the emergency department revealed thyroid-stimulating hormone (TSH; 679 μ/mL; normal 0.35–5.5 μ/mL), free thyroxine (T_4; 0.36 ng/dL; normal 0.93–1.7 ng/dL), triiodothyronine (T_3; 23.53 ng/dL; normal 84–202 ng/dL), calcium (9.8 mg/dL), potassium (K; 3.7 mmol/L), and negative urine toxicology. Corrected QT interval (QTc) was noted to be 523 ms on admission electrocardiography, and the patient was consequently started on intravenous hydrocortisone and synthroid therapy. During the first day of hospitalization, patient had pre-syncope with corresponding telemetry strip showing torsades de pointes (*Figure 1*). Patient was given 2 g of intravenous magnesium sulfate and transferred to the coronary care unit. Echocardiography revealed preserved left ventricular ejection fraction. QTc improved to 329 ms after regaining euthyroid status with corresponding TSH 0.3 (μ/mL), and the decision to place an implantable cardioverter–defibrillator (ICD) was deferred. The patient was later tested for a congenital long QT syndrome in a panel of 30 genes accounting for most of the arrhythmic hereditary diseases. Genetic testing revealed heterozygosity for a novel frameshift mutation c.2470delG in *KCNH2* (*HERG*) gene. Although the c2470delG mutation in the *KCNH2* gene has not been reported to our knowledge, this mutation causes a shift in the reading frame starting at codon alanine 824, changing it to a proline, and creating a premature stop codon at Position 44 of the new reading frame, denoted p.Ala824ProfsX44. This mutation is expected to result in either an abnormal truncated protein product or a loss of protein from this allele through non-sense-mediated messenger RNA decay. The patient was also heterozygous for a published variant of unknown significance in the *GPD1L* gene.[1]



Serum TSH correlates with cardiovascular changes and risk of potentially life-threatening arrhythmias in hypothyroid patients. Modulation of K channels function by low circulating T_3 leads to a disproportionate lengthening of the action potential and consequent lengthening of QT interval.[2] Ventricular arrhythmias are usually reversible with levothyroxine replacement therapy, unwarranting ICD placement following acute-phase resolution.[2,3]

Conflict of interest: none declared.

References

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